CHAPTER 8

Lens

Introduction

The crystalline lens is a biconvex, avascular, transparent structure enclosed by a capsule, a basement membrane secreted by the lens epithelium. The capsule, responsible for moulding the lens substance during accommodation, is thickest in the equatorial zone and thinnest at the posterior pole of the lens. A ring of zonular fibres, which insert in the equatorial region, suspends the lens from the ciliary body. A monolayer of epithelium lines only the anterior and equatorial lens capsule. Cells in the equatorial region exhibit mitotic activity. Newly formed epithelial cells elongate to form fibres, which lose their organelles, thus optimizing lens transparency. Lens substance may be conceptualized as consisting of the nucleus, the central compacted core, which is surrounded by the cortex. New lens fibres are continuously laid down subcapsularly throughout life, resulting in the older layers acquiring progressively deeper localizations within the lens substance. The lens thus grows, in both anteroposterior and equatorial dimensions, throughout life. The normal lens is transparent; any congenital or acquired opacity in the lens capsule or substance, irrespective of the effect on vision, is a cataract.

Acquired cataract

Age-related cataract

Morphological classification

1. Subcapsular cataract

a. *Anterior subcapsular* cataract lies directly under the lens capsule and is associated with fibrous metaplasia of the lens epithelium (Fig. 8.1).

b. *Posterior subcapsular* cataract lies just in front of the posterior capsule and manifests a vacuolated, granular or plaque-like appearance (Fig. 8.2). Due to its location at the nodal point of the eye, a posterior subcapsular opacity has a more profound effect on vision than a comparable nuclear or cortical cataract. Patients are particularly troubled under conditions of miosis, such as produced by headlights of oncoming cars and bright sunlight. Near vision is also frequently impaired more than distance vision.

2. **Nuclear** cataract starts as an exaggeration of the normal ageing changes involving the lens nucleus (Fig. 8.3). It is often associated with myopia due to an increase in the refractive index of the lens nucleus and also with increased spherical aberration. Some elderly patients may consequently be able to read again without spectacles ('second sight of the aged'). Nuclear sclerosis is characterized in its early stages by a yellowish hue due to the deposition of urochrome pigment (Fig. 8.4). When advanced the nucleus appears brown (a brunescent cataract) (Fig. 8.5). Such cataracts are of hard consistency, which is surgically relevant.

3. **Cortical** cataract may involve the anterior, posterior or equatorial cortex. The opacities start as clefts (Fig. 8.6)



Fig. 8.2
Posterior subcapsular cataract



Fig. 8.1
Anterior subcapsular cataract



Fig. 8.3
Early nuclear cataract


Fig. 8.4
Moderate nuclear cataract

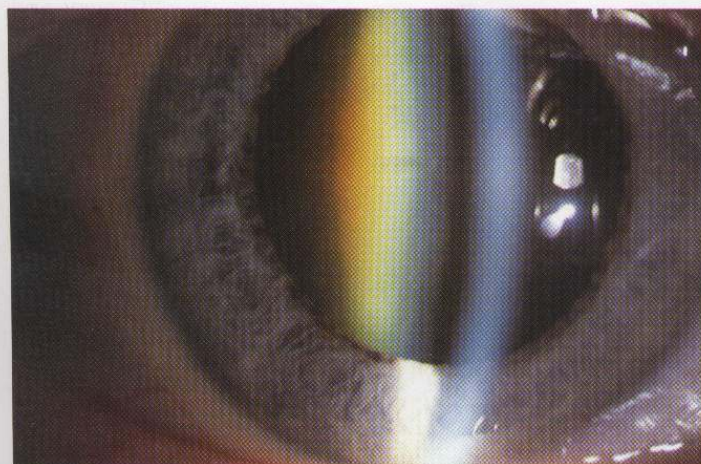

Fig. 8.5
Brunescent nuclear cataract


Fig. 8.6
Cortical clefts

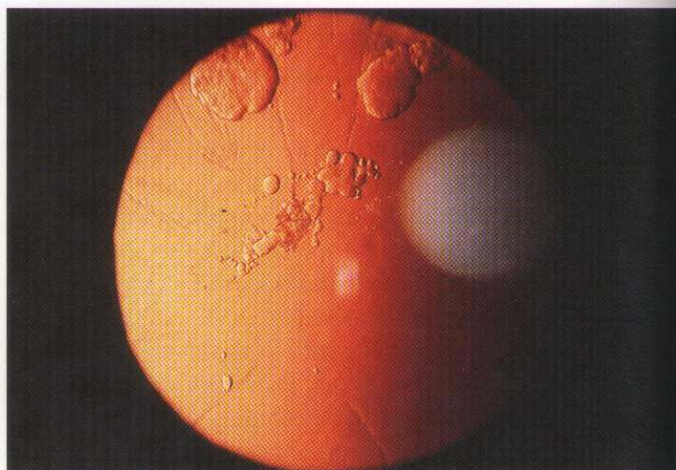
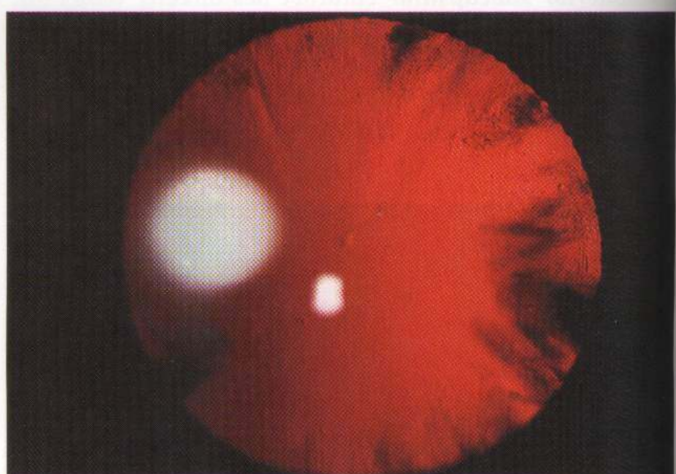

Fig. 8.7
Cortical vacuoles


Fig. 8.8
Advanced cortical cataract


Fig. 8.9
Cortical cataract seen on retroillumination

and vacuoles (Fig. 8.7) between lens fibres due to hydration of the cortex. Subsequent opacification results in typical cuneiform (wedge-shaped) or radial spoke-like opacities, often initially in the inferonasal quadrant. Both cortical and subcapsular cataracts are white on oblique illumination (Fig. 8.8) and appear black, silhouetted against the red reflex, on retroillumination (Fig. 8.9).

4. **Christmas tree** cataract, which is uncommon, is characterized by striking, polychromatic, needle-like deposits in the deep cortex and nucleus which may be solitary (Fig. 8.10) or associated with other opacities (Fig. 8.11).

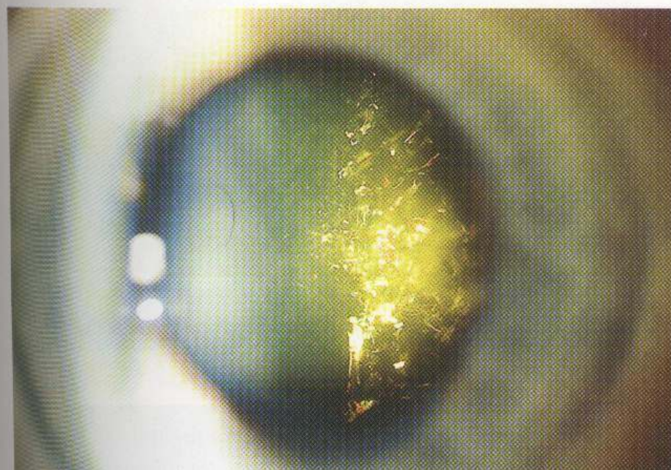


Fig. 8.10
Christmas tree cataract

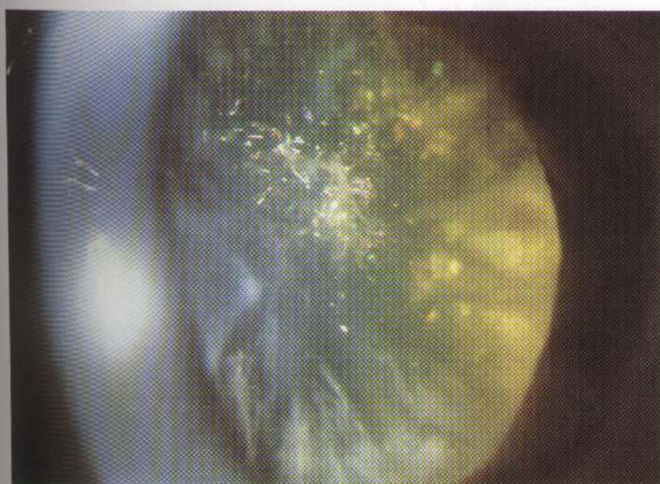


Fig. 8.11
Mixed christmas tree and cortical cataract



Fig. 8.12
Mature cataract

Classification according to maturity

1. **An immature** cataract is one in which the lens is partially opaque.
2. **A mature** cataract is one in which the lens is completely opaque (Fig. 8.12).
3. **A hypermature** cataract has a shrunken and wrinkled anterior capsule due to leakage of water out of the lens (Fig. 8.13).
4. **A morgagnian** cataract is a hypermature cataract in which total liquefaction of the cortex has allowed the nucleus to sink inferiorly (Fig. 8.14).

Presenile cataract

Cataract may develop early in the following conditions:

1. **Diabetes mellitus** (*see* Chapter 20), in addition to causing cataract, can affect the refractive index of the lens and also its amplitude of accommodation.
   a. *Classical diabetic* cataract is rare. Hyperglycaemia is reflected in a high level of glucose in the aqueous

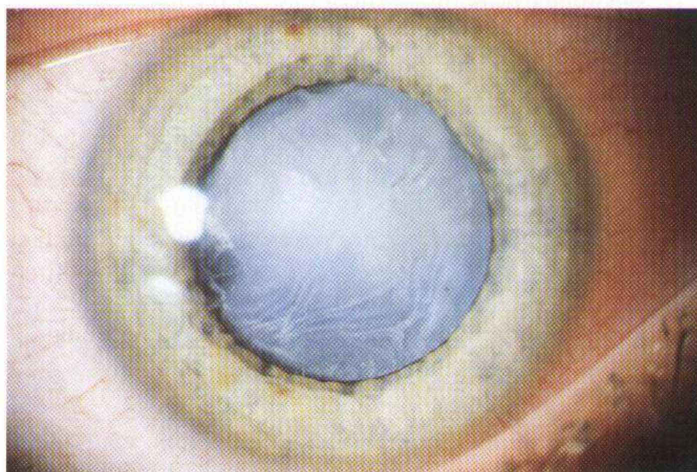


Fig. 8.13
Hypermature cataract with wrinkling of the anterior capsule

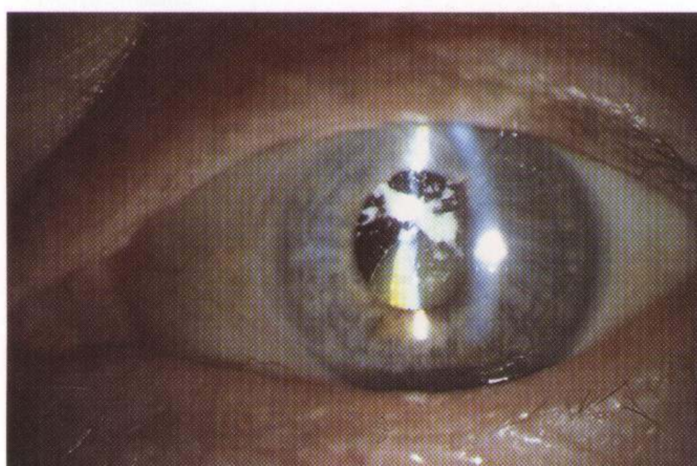


Fig. 8.14
Morgagnian cataract

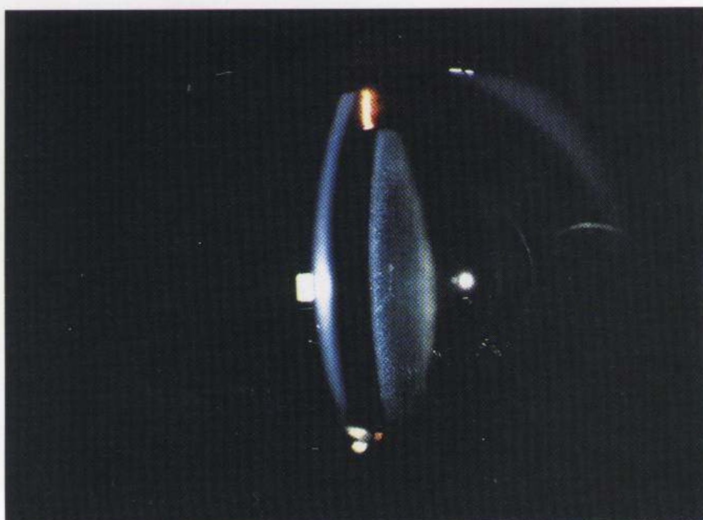


Fig. 8.15
Diabetic snowflake cataract (Courtesy of A. Fielder)

humour, which diffuses into the lens. Here glucose is metabolized by aldose reductase into sorbitol, which then accumulates within the lens, resulting in secondary osmotic overhydration of the lens substance. In mild degree, this may affect the refractive index of the lens with consequent fluctuation of refraction *pari passu* with the plasma glucose level (hyperglycaemia resulting in myopia). Cortical fluid vacuoles develop and later evolve into frank opacities. Classical diabetic cataract consists of snowflake cortical opacities (Fig. 8.15) occurring in the young diabetic. Such a cataract may resolve spontaneously or mature within a few days.

b. Age-related cataract occurs earlier in diabetes mellitus. Nuclear opacities are common and tend to progress rapidly.

c. Premature presbyopia may be seen due to reduced pliability of the lens.

2. **Myotonic dystrophy** (*see* Chapter 20). About 90% of patients develop visually innocuous, fine cortical iridescent opacities in the third decade, which evolve into visually disabling stellate posterior subcapsular cataract by the fifth decade (Fig. 8.16). Occasionally cataract may antedate myotonia.

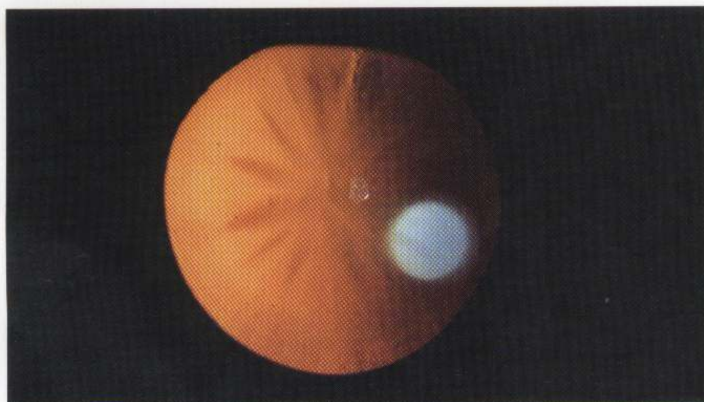


Fig. 8.16
Stellate posterior subcapsular cataract in myotonic dystrophy seen on retroillumination

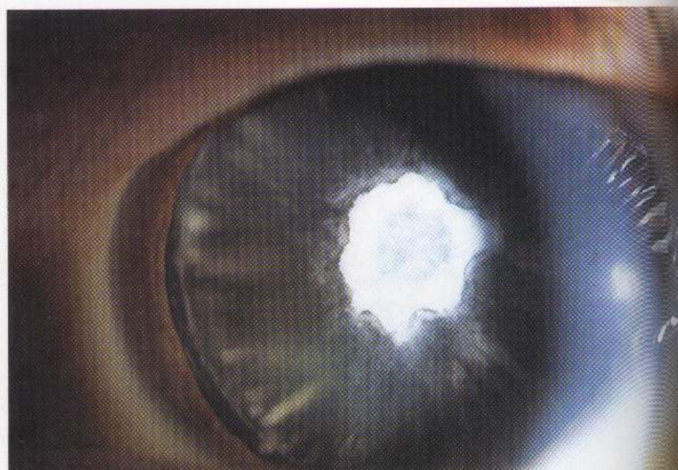


Fig. 8.17
Shield-like anterior subcapsular cataract in atopic dermatitis

3. **Atopic dermatitis** (*see* Chapter 20). About 10% of patients with severe atopic dermatitis develop cataracts in the second to fourth decades. The opacities are often bilateral and may mature quickly.
   a. Shield-like dense anterior subcapsular plaque which wrinkles the anterior capsule is characteristic (Fig. 8.17).
   b. Posterior subcapsular opacities resembling a complicated cataract may also occur.
4. **Neurofibromatosis type 2** (*see* Chapter 20) is associated with posterior subcapsular or posterior cortical opacities.

Traumatic cataract

Trauma is the most common cause of unilateral cataract in young individuals. The following may be responsible:

1. **Direct penetrating** injury to the lens (Fig. 8.18).
2. **Concussion** may cause an 'imprinting' of iris pigment on the anterior lens capsule (Vossius ring) (Fig. 8.19) as well

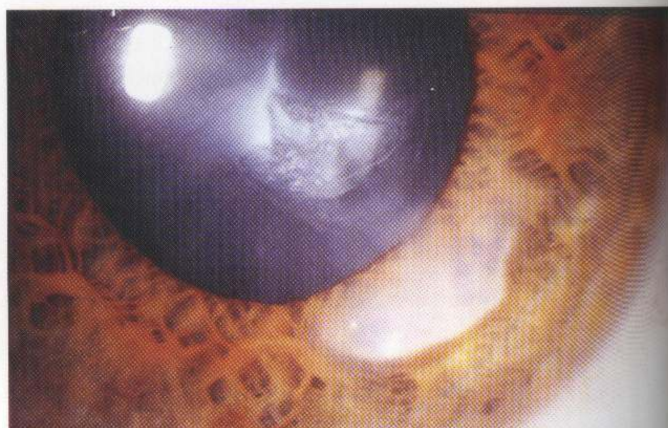


Fig. 8.18
Cataract caused by penetrating trauma

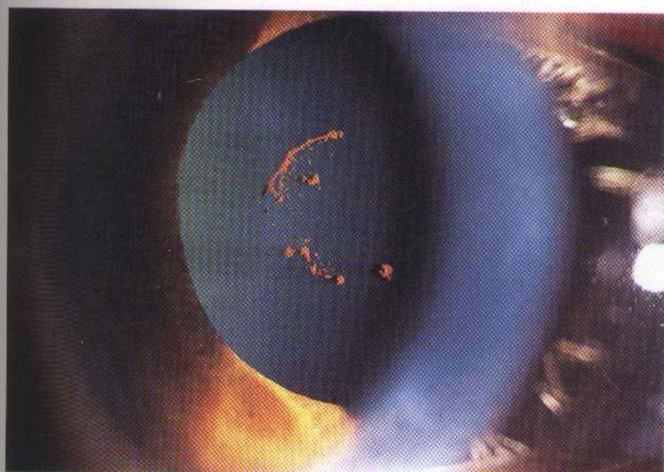

Fig. 8.19
Vossius ring due to blunt trauma

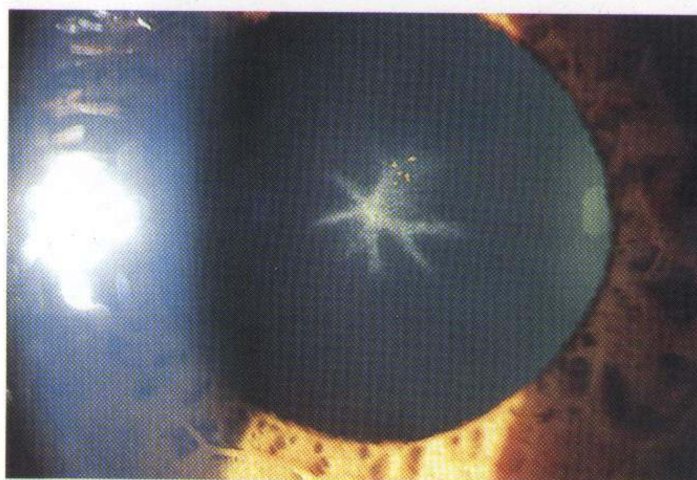

Fig. 8.20
Flower-shaped (rosette) cataract due to blunt trauma

as striking flower-shaped cortical opacities (rosette cataract) (Fig. 8.20).

3. **Electric shock** and **lightning** are rare causes.
4. **Ionizing radiation** to ocular tumours.
5. **Infrared radiation**, if intense as in glassblowers, may rarely cause true exfoliation or lamellar delamination of the anterior lens capsule in which the superficial portion of a thickened capsule splits from the deeper layer and extends into the anterior chamber. This condition is distinct from pseudoexfoliation in which fibrillary material is deposited on the anterior lens surface and other ocular structures (*see* Chapter 9).

Drug-induced cataract

1. **Steroids**, both systemic and topical, are cataractogenic. The lens opacities are initially posterior subcapsular; later the anterior subcapsular region becomes affected. The relationship between weekly systemic dose, duration of administration, total dose and cataract formation is unclear. It is thought that patients on less than 10 mg prednisolone (or equivalent), or treated for less than 4 years, may be immune. Although it is believed that children may be more susceptible to the cataractogenic effects of systemic steroids, individual (genetic) susceptibility may also be of relevance. It has therefore been suggested that the concept of a safe dose be abandoned. Patients who develop lens changes should have their dosage reduced to a minimum consistent with control of the underlying disease, and if possible be considered for alternate-day therapy. Early opacities may regress if therapy is discontinued; alternatively progression may occur despite withdrawal and warrant surgical intervention.
2. **Chlorpromazine** may cause the deposition of innocuous, fine, stellate, yellowish-brown granules on the anterior lens capsule within the pupillary area (Fig. 8.21). Diffuse, granular deposits on the corneal endothelium and in the deep stroma may also occur. Both lenticular and corneal deposits are dose-related and usually irreversible. In very high doses (>2400 mg daily) this drug may cause retinotoxicity (*see* Chapter 13).
3. **Busulphan** (Myleran), used in the treatment of chronic myeloid leukaemia, may occasionally cause lens opacities.


Fig. 8.21
Anterior lens deposits due to the long-term use of chlorpromazine

4. **Amiodarone**, used in the treatment of cardiac arrhythmias, causes visually inconsequential anterior subcapsular lens deposits in about 50% of patients on moderate to high doses. Vortex keratopathy may also occur (*see* Chapter 5).
5. **Gold**, used in the treatment of rheumatoid arthritis, causes innocuous anterior capsular deposits in about 50% of patients on treatment for longer than 3 years.
6. **Allopurinol**, used in the treatment of hyperuricaemia and chronic gout, increases the risk of cataract formation in elderly patients, if the cumulative dose exceeds 400 g or duration of administration exceeds 3 years.

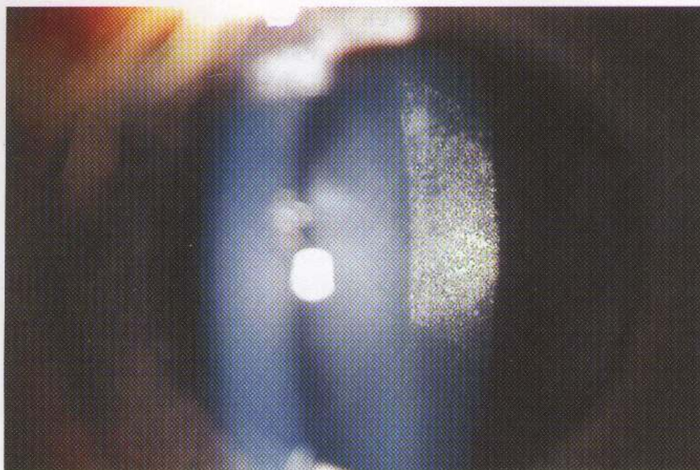


Fig. 8.22
Posterior polychromatic lustre in chronic anterior uveitis

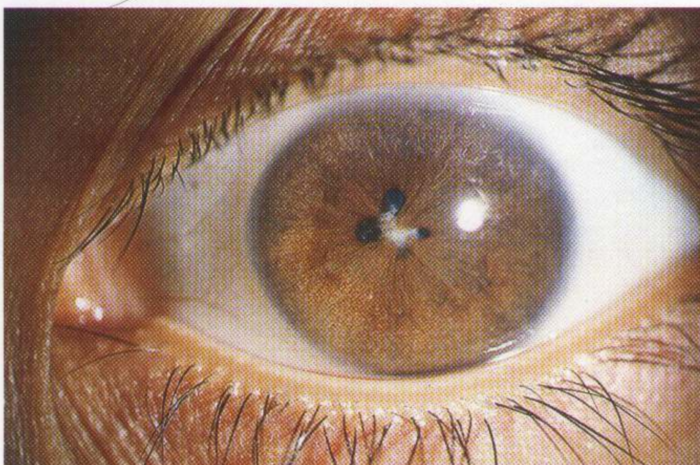


Fig. 8.23
Anterior cataract and extensive posterior synechiae in chronic anterior uveitis



Fig. 8.24
Mature cataract and posterior synechiae in chronic anterior uveitis

Secondary cataract

A secondary (complicated) cataract develops as a result of some other primary ocular disease.

1. **Chronic anterior uveitis** is the most common cause of secondary cataract. The earliest finding is a polychromatic lustre at the posterior pole of the lens (Fig. 8.22) which may not progress if the uveitis is arrested. If the inflammation persists, posterior and anterior opacities develop (Fig. 8.23) and may progress to maturity (Fig. 8.24). Lens opacities appear to progress more rapidly in the presence of posterior synechiae.

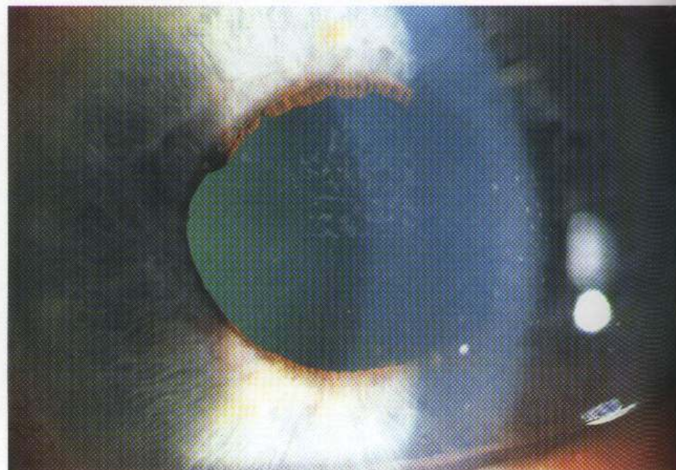


Fig. 8.25
Anterior capsular cataract following acute angle-closure glaucoma (glaukomflecken)

2. **Acute congestive angle-closure glaucoma** may cause small, grey-white, anterior, subcapsular or capsular opacities within the pupillary area (glaukomflecken) (Fig. 8.25). They represent focal infarcts of the lens epithelium and are pathognomonic of past acute angle-closure glaucoma.
3. **High (pathological) myopia** is associated with posterior subcapsular lens opacities and early-onset nuclear sclerosis, which may ironically increase the myopic refractive error. Simple myopia, however, is not associated with such cataract formation.
4. **Hereditary fundus dystrophies** such as retinitis pigmentosa, Leber congenital amaurosis, gyrate atrophy and Stickler syndrome may be associated with posterior subcapsular lens opacities (*see* Chapter 15). Cataract surgery may occasionally improve visual acuity even in the presence of severe retinal changes.

Management of age-related cataract

Introduction

Indications for surgery

1. **Visual improvement** is by far the most common indication for cataract surgery, although requirements vary from person to person. Surgery is indicated only if and when cataract develops to a degree sufficient to cause difficulty in performing daily essential activities. If the

patient desires to drive or continue a specific occupation, visual function below legally prescribed levels may necessitate cataract surgery.

2. **Medical indications** are those in which a cataract is adversely affecting the health of the eye, for example, phacolytic glaucoma or phacomorphic glaucoma (*see* Chapter 9). Cataract surgery to improve the clarity of the ocular media may also be required in the context of fundal pathology (e.g. diabetic retinopathy) requiring monitoring or treatment with laser photocoagulation.
3. **Cosmetic indications** are rare, such as when a mature cataract in an otherwise blind eye is removed to restore a black pupil.

Preoperative evaluation

Apart from a general medical examination, a patient due to undergo cataract surgery requires a detailed and pertinent ophthalmic examination, with special regard to the following:

1. **Cover test**. A heterotropia may indicate amblyopia, which carries a guarded visual prognosis, or the possibility of diplopia if the vision is improved.
2. **Pupillary responses**. Because a cataract never produces an afferent pupillary defect, its presence implies additional pathology, which may influence the final visual outcome.
3. **Ocular adnexa**. Dacryocystitis, blepharitis, chronic conjunctivitis, lagophthalmos, ectropion, entropion and tear film abnormalities may predispose to endophthalmitis and require effective preoperative treatment.
4. **Cornea**. A wide arcus senilis or stromal opacities may prejudice a good surgical view. Guttata indicate endothelial dysfunction and consequent vulnerability to decompensation secondary to operative trauma.
5. **Anterior segment**. A shallow anterior chamber can render cataract surgery difficult. Pseudoexfoliation indicates a weak zonule, with the possibility of problems during surgery. A poorly dilating pupil can make cataract surgery difficult. Recognition of this allows intensive preoperative mydriatic drops or planned stretching of the pupil prior to capsulorhexis. A poor red reflex compromises the performance of a good capsulorhexis. This can be overcome by staining the capsule with a dye such as trypan blue.
6. **Lens**. The type of cataract is relevant. Nuclear cataracts tend to be hard and require more phaco power, while cortical and subcortical cataracts tend to be softer.
7. **Intraocular pressure**. Any glaucoma or ocular hypertension must be noted.
8. **Fundus**. Recognition of fundal pathology such as age-related macular degeneration, which may affect the visual outcome.

Biometry

Surgical removal of the crystalline lens subtracts approximately 20 D from the refracting system of the eye. The aphakic eye is grossly hypermetropic; modern cataract surgery therefore involves the implantation of an intraocular lens (IOL), ideally in the same location as the surgically removed crystalline lens. Biometry affords calculation of the lens power likely to result in emmetropia or, alternatively, a desired postoperative refractive error. In its simplest form, biometry involves two ocular parameters: (a) *keratometry*—the curvature of the anterior corneal surface (steepest and flattest meridians), expressed in dioptres or millimetres of radius of curvature and (b) *axial length*—the anteroposterior dimension of the eye in millimetres, measured on A-scan ultrasonography.

1. **SRK formula**. Perhaps the most commonly used mathematical formula to calculate IOL power is that developed by Sanders, Retzlaff and Kraff which states that $P = A - 2.5L - 0.9K$
   - P is the lens power required to generate postoperative emmetropia.
   - A is the A constant, which varies (between 114 and 119) with different IOLs.
   - L is the axial length in millimetres.
   - K is the average keratometry reading in dioptres.

   Numerous other formulae, incorporating additional parameters such as anterior chamber depth and individualized surgeon factors have been developed to optimize the accuracy of preoperative prediction.
2. **Postoperative refraction**. Emmetropia is perhaps the ideal postoperative refraction, with spectacles needed only for close work (since an IOL cannot accommodate). In practice, most surgeons aim for a small degree of myopia (about 0.25 D) to offset possible error in biometry. This is because a slight degree of myopia is acceptable in most patients, and may even be advantageous, while postoperative hypermetropia, which necessitates spectacles for clear vision at all distances, is poorly tolerated. The planning of postoperative refraction also needs to take account of the other eye. If the other eye has clear vision with a significant refractive error and is unlikely to require surgery, then postoperative refraction should be targeted at within 2 D of the other eye, to avoid problems with binocular coordination.

Anaesthesia

Evidence for the benefit of local anaesthesia (LA) over general anaesthesia (GA) for most intraocular procedures is sparse. The choice is generally determined by patient preference and the clinical judgement of the surgical team. Day-case cataract surgery under LA is safe and generally preferred by patients and staff alike. It affords significant economic benefits and is the option of choice.

1. **Retrobulbar block** is given into the muscle cone behind the globe close to the ciliary ganglion with a 1.5 inch (38 mm) needle. It also provides akinesia so that ocular movements are greatly decreased or eliminated altogether. Retrobulbar injection requires considerable skill and experience and is occasionally associated with serious

complications such as orbital haemorrhage, penetration of the globe, intravascular injection, damage to the optic nerve and brain stem anaesthesia. Temporary problems include ptosis and diplopia. Retrobulbar anaesthesia often requires a separate facial block to paralyse the orbicularis oculi.

2. **Peribulbar block** is given through the skin or conjunctiva with a 1 inch (25 mm) needle. More than one injection and a greater volume of anaesthetic agent is required compared with retrobulbar block. Because the needle is shorter the risk of brain stem anaesthesia is reduced although haemorrhage and ocular penetration may occur.
3. **Parabulbar (sub-Tenon) block** involves passing a blunt-tipped cannula through an incision in the conjunctiva and Tenon capsule 5 mm from the limbus, and along the sub-Tenon space. The anaesthetic is injected beyond the equator of the globe. Although anaesthesia is good and complications minimal, akinesia is variable.
4. **Topical–intracameral anaesthesia** involves initial surface anaesthesia with drops or gel (proxymetacaine 0.5%, lignocaine 4%) and intracameral injection or infusion of diluted preservative-free anaesthetic.

Intraocular lenses

Basic aspects

1. **Positioning**. An IOL consists of the optic (the central refracting element) and the haptics, which sit in contact with the ocular structures (capsular bag, ciliary sulcus or anterior chamber angle), thus affording optimal and stable position (centration) of the optic. Modern cataract surgery, with preservation of the capsular 'bag', affords positioning of the IOL in the ideal location—'in the bag'. Complicated surgery, with rupture of the posterior capsule, may, however, necessitate alternative positioning of the IOL, in the posterior chamber (with haptics in the ciliary sulcus) or in the anterior chamber with the haptics supported in the chamber angle. The latter is designated an AC-IOL in contrast to the former two, which are PC-IOLs.
2. **Designs** are numerous and continue to evolve. The lenses may be rigid or flexible. A rigid IOL requires an incision larger than the diameter of the optic, often 5–6.6 mm, for insertion. A flexible IOL, however, may be folded with forceps or loaded into an injector/delivery system and inserted through a much smaller incision, often 2.5–3 mm. Haptics are made from polymethylmethacrylate (PMMA), polypropylene (Proline) or polyamide and may be in the form of loops or plates. In a one-piece IOL the haptics and optic are made from the same material and have no joints; a three-piece IOL is characterized by optics and haptics from different materials, which necessarily are joined together. Optics may also be of different sizes and shapes. Conventional IOLs are monofocal; lately multifocal designs that allow clear vision at different distances are being developed.
3. **Rigid IOLs** are made entirely from PMMA. The composition of PMMA varies depending on the manufacturing process. Compression-moulded and lathe-cut IOLs require high-molecular-weight PMMA, while injection-moulded lenses need lower-molecular-weight PMMA. Modern rigid IOLs are one-piece to facilitate maximal stability and fixation.
4. **Foldable IOLs** are made from the following materials:
   a. ***Silicone*** IOLs, both three-piece loop and one-piece plate haptics, are associated with lower rates of posterior capsular opacification than PMMA lenses.
   b. ***Acrylic*** IOLs, three-piece or one-piece, may be hydrophobic (water content <1%) or hydrophilic (water content 18–35%). Certain acrylic IOLs inhibit opacification of the posterior capsule.
   c. ***Hydrogel*** IOLs are similar to hydrophylic acrylic IOLs, have a high water content (38%) and are available only as three-piece designs.
   d. ***Collamer*** IOLs, made from a mixture of collagen and hydrogel, have recently been introduced.

Cataract surgery

Principles

1. **Extracapsular cataract extraction** (ECCE) requires a relatively large circumferential limbal incision (8–10 mm) through which the lens nucleus is extracted and the cortical matter aspirated, leaving behind an intact posterior capsule. The IOL is then inserted.
2. **Phacoemulsification** (phaco) has become the preferred method of cataract extraction over the last decade. A small hollow needle, usually titanium, attached to a handpiece containing a piezo-electric crystal, vibrates longitudinally at ultrasonic frequencies. The tip is applied to the lens nucleus; cavitation occurs at the tip as the nucleus is emulsified; an irrigating/aspiration system removes this emulsified material from the eye. The IOL is then inserted (if folded) or injected through a much smaller incision than in ECCE. The smaller incision renders the operation safer, since decompression of the eye is avoided; this reduces the incidence of operative complications such as suprachoroidal haemorrhage, shallowing of the anterior chamber and vitreous prolapse in the event of rupture of the posterior capsule. In addition the procedure is associated with little postoperative astigmatism and early stabilization of refraction (usually 3 weeks). Postoperative wound-related problems such as iris prolapse are almost eliminated.

Technique of extracapsular extraction

1. A circumferential vertical partial thickness incision is made in the peripheral clear cornea at the limbus and the anterior chamber entered with a keratome.
2. Viscoelastic substance such as sodium hyaluronate or hydroxymethylpropylcellulose is injected into the anterior chamber. This maintains the chamber and protects the corneal endothelium.

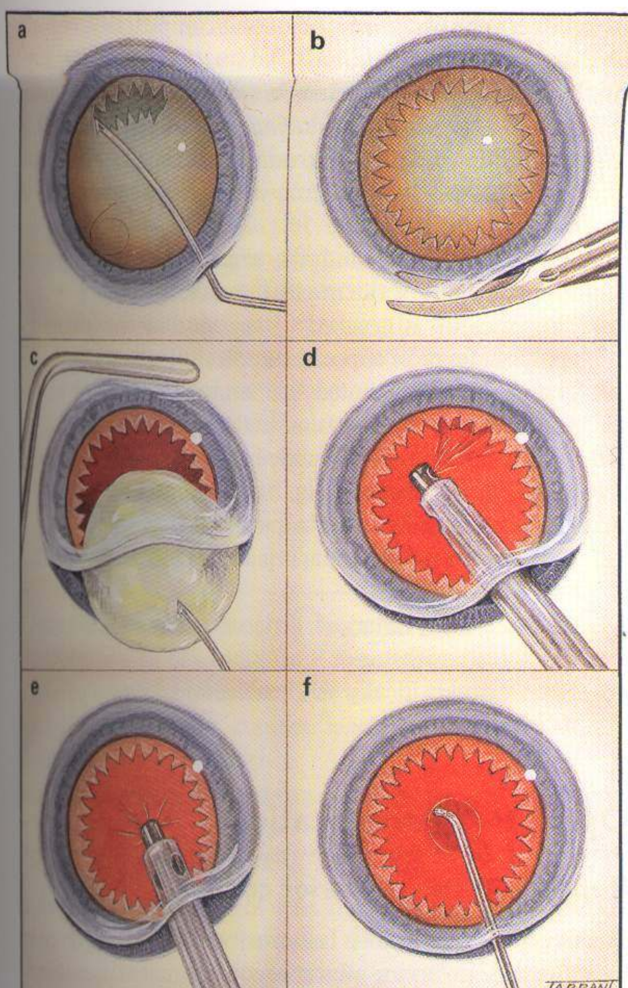


Fig. 8.26
Extracapsular cataract extraction (*see text*)

3. The cystitome is introduced into the anterior chamber and multiple small radial cuts are made in the anterior capsule for 360° (Fig. 8.26a). These cuts are then joined together to create a 'can-opener' capsulotomy. Alternatively, a capsulorhexis may be performed, which involves making a controlled circular tear in the capsule.
4. The limbal partial-thickness incision is completed with scissors (Fig. 8.26b).
5. Hydrodissection is performed by injecting balanced salt solution (BSS) with a special blunt-tipped (Rycroft) cannula between the edge of the capsule and the peripheral lens cortex in order to free the lens matter from the capsular bag.
6. The nucleus is expressed by applying pressure alternatively at the superior and inferior limbus, (Fig. 8.26c) or by manipulating it out with a vectis.
7. The tip of the infusion–aspiration cannula is introduced into the anterior chamber and passed under the lens capsule at 6 o'clock. Strands of cortex are engaged into the port by activating the suction mechanism (Fig. 8.26d).
8. The cortex is then dragged centrally and aspirated under direct visualization. This manoeuvre is repeated sequentially until all cortex has been removed. It is important not to inadvertently aspirate the posterior capsule because this may cause it to rupture, with numerous attendant complications. Aspiration of the capsule is indicated by the appearance of fine sharp lines radiating from the aspiration port (Fig. 8.26e). Aspiration should be immediately terminated and fluid reflux activated to disengage the capsule.
9. If necessary, the posterior capsule may be polished to remove any small residual subcapsular plaques (Fig. 8.26f).
10. Viscoelastic substance is injected into the capsular bag to facilitate subsequent insertion of the IOL (Fig. 8.27a).
11. The IOL is grasped by the optic and its anterior surface coated with viscoelastic substance (Fig. 8.27b).
12. The leading haptic is inserted through the lips of the incision and then passed under the lens capsule at 6 o'clock (Fig. 8.27c).
13. The tip of the superior haptic is grasped with forceps and advanced into the anterior chamber (Fig. 8.27d). As the superior pole of the haptic clears the edge of the pupil, the arm is pronated to ensure that on release the haptic will spring open under the iris and not out of the incision.

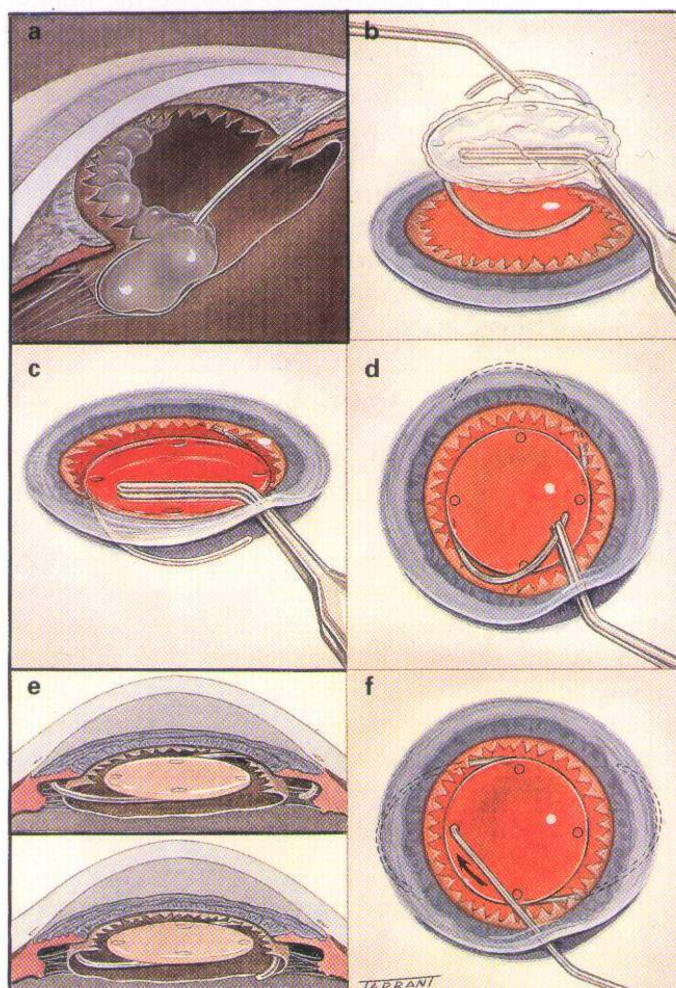


Fig. 8.27
Extracapsular cataract extraction (*see text*)

> ***NB:*** Both haptics should preferably be placed in the capsular bag (Fig. 8.27e, bottom) rather than in the ciliary sulcus (Fig. 8.27e, top).

14. The IOL is dialled into the horizontal position by engaging the guide holes with a special hook (Fig. 8.27f).
15. The pupil is constricted by injecting Miochol (acetylcholine) into the anterior chamber, viscoelastic substance is aspirated, and the incision is sutured.

Phacoemulsification

The technique is constantly changing and there are many variations. The basic steps for a classic technique (4 quadrant 'divide and conquer') are as follows:

1. A self-sealing tunnel incision is made into the anterior chamber. This may be in clear peripheral cornea, preferably temporally, or via a scleral tunnel incision, usually superiorly.
2. Viscoelastic substance is injected into the anterior chamber.
3. A second stab incision is made in clear peripheral cornea, at right angles to the first incision.
4. A continuous capsulorhexis is performed (Fig. 8.28a).
5. Hydrodissection is performed to mobilize the nucleus (Fig. 8.28b). A retrocortical 'fluid wave' over the red reflex can be observed, indicating completeness of hydrodissection.
6. 'Sculpting' of the nucleus is performed with the phaco probe to create a groove. The nucleus is rotated with a second instrument introduced through the second incision and a second groove is made at right angles to the first (Fig. 8.28c).
7. The phaco probe and the second instrument engage opposite walls of the nuclear groove.
8. The nucleus is cracked in the base of the groove by applying force in opposite directions (Fig. 8.28d).
9. The nucleus is rotated 90° and a crack made in the perpendicular furrow in a similar manner.
10. Each quadrant of the nucleus is fragmented, emulsified and aspirated in turn (Fig. 8.28e).
11. The remaining cortex is aspirated (Fig. 8.28f).
12. Viscoelastic substance is injected to inflate the capsular bag.
13. The incision is enlarged, if necessary, and IOL inserted.
14. Viscoelastic substance is aspirated.
15. The wound is ideally self-healing and does not require a suture.

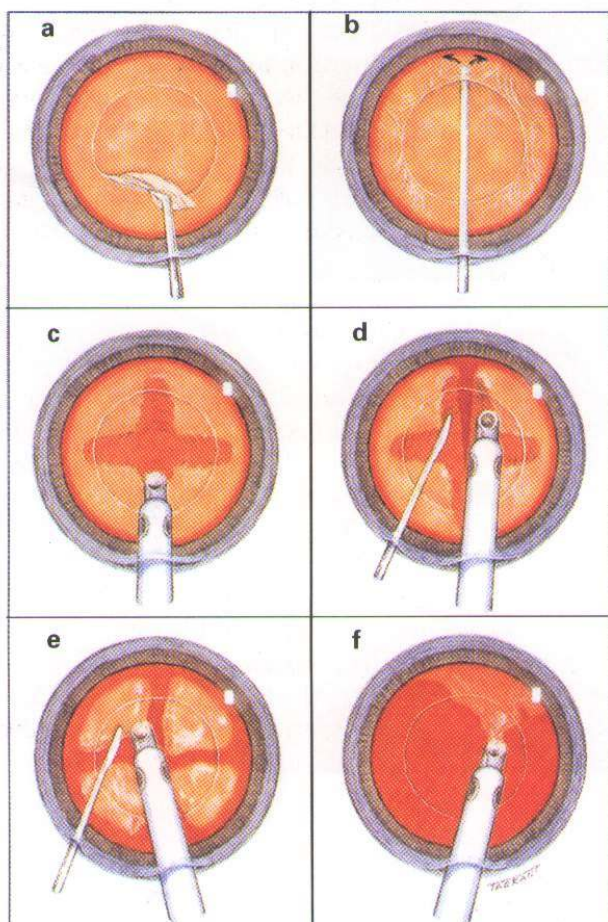


Fig. 8.28
Phacoemulsification (see *text*)

Operative complications

Rupture of the posterior capsule

This is potentially serious because it may be accompanied by vitreous loss, posterior migration of lens material and rarely expulsive haemorrhage. Long-term complications of vitreous loss, particularly if inappropriately managed, include updrawn pupil, uveitis, vitreous touch, vitreous wick syndrome, secondary glaucoma, posterior dislocation of the IOL, retinal detachment and chronic cystoid macular oedema.

1. Signs
- Sudden deepening of the anterior chamber and momentary pupillary dilatation.
- The nucleus falls away and will not come towards the phaco tip.
- Vitreous may be aspirated into the phaco tip.
- The torn capsule or vitreous may be directly visible.

2. Management depends on the point in the procedure at which the tear occurred, the magnitude of the tear and the presence or absence of vitreous prolapse. The main principles of management are as follows:

a. Viscoelastic substance may be injected behind nuclear material with the purpose of expressing it into the anterior chamber and also preventing anterior herniation of vitreous.
b. A lens glide is passed behind the lens fragments to cover the capsular defect.
c. The lens fragments are expressed by visco-expression or removed by phaco.
d. All vitreous must be removed from the anterior chamber and the wound with a vitrector.
e. A decision has to be taken whether or not to implant an IOL based on the following criteria:

- If there is significant loss of lens material into the vitreous cavity, an IOL should not be implanted, since it may compromise visualization of the fundus, vital to successful pars plana vitrectomy (*see* later). IOL implantation can be performed at the time of vitrectomy.
- A very small posterior capsular tear may allow careful in-the-bag implantation of a PC-IOL.
- A large tear may still allow ciliary sulcus fixation of a PC-IOL with the optic captured in the capsular bag, especially if the anterior capsulorhexis is intact.
- Insufficient capsular support may necessitate that the IOL be sutured to the sulcus or alternatively, that an AC-IOL be implanted with the aid of a glide (Fig. 8.29). AC-IOLs are, however, associated with a higher risk of complications including bullous keratopathy, hyphaema, iris tuck and pupillary irregularities.

> *NB:* Conditions may not be favourable for correct positioning even of an AC-IOL. It may then be safer not to proceed with lens implantation and to consider visual rehabilitation with a contact lens or secondary IOL implantation at a later date.

Posterior loss of lens fragments

Dislocation of fragments of lens material into the vitreous cavity (Fig. 8.30) after zonular dehiscence or posterior capsule rupture is rare but potentially serious because it may result in glaucoma, chronic uveitis, retinal detachment and chronic cystoid macular oedema. This complication is more commonly associated with phaco than ECCE. Initially, any uveitis or raised intraocular pressure must be treated. The patient should then be referred to a vitreoretinal surgeon for removal of nuclear fragments by pars plana vitrectomy.

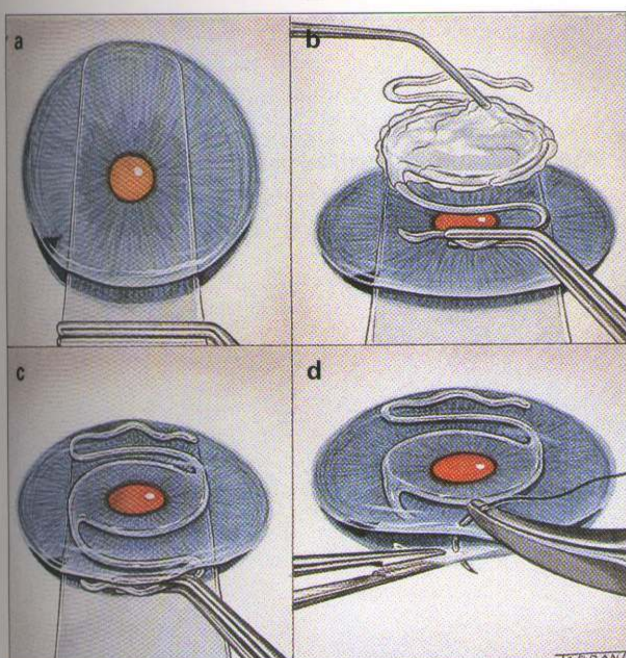


Fig. 8.29
Insertion of an anterior chamber intraocular lens

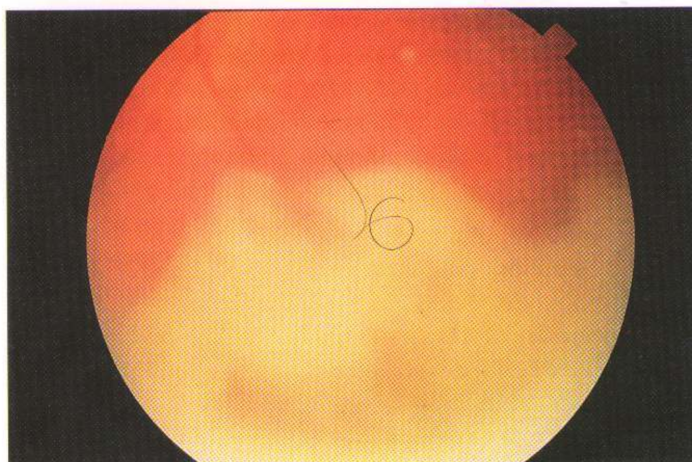


Fig. 8.30
Nuclear material in the vitreous

1. **Timing of surgery** is controversial. Some suggest the fragments be removed within the first week as later removal is associated with poorer visual outcome. Others recommend that surgery should be deferred for 2–3 weeks provided uveitis and raised intraocular pressure are controlled medically. This allows the lens material to become hydrated and softer, facilitating removal with a vitreous cutter alone (*see* below).
2. **Surgical technique** involves pars plana vitrectomy and removal of soft fragments with a vitreous cutter. Hard nuclear fragments are floated clear of the retina with heavy liquids (i.e. perfluorocarbons) and either emulsified with the fragmatome in the mid-vitreous cavity or delivered via a corneal incision or scleral pocket. An alternative method of removing hard nuclear material involves crushing them between the illumination probe and the vitreous cutter and then 'force feeding' them into the cutting port.

Posterior dislocation of IOL

Dislocation of a PC-IOL into the vitreous cavity reflects inappropriate implantation and is rare but serious. If the IOL is left it may lead to vitreous haemorrhage, retinal detachment, uveitis and chronic cystoid macular oedema. Treatment involves pars plana vitrectomy with removal, repositioning or exchange of the IOL. If adequate capsular support is present the same IOL may be repositioned into the ciliary sulcus. Inadequate capsular support affords the following alternatives: (a) removing the IOL and leaving the eye aphakic, (b) removing the IOL and replacing it with an AC-IOL, (c) scleral fixation of the existing IOL with a non-absorbable suture and (d) using an iris-fixated IOL.

Suprachoroidal haemorrhage

This is a bleed into the suprachoroidal space which may result in extrusion of intraocular contents (expulsive haemorrhage) or apposition of retinal surfaces. It is a dreaded but rare complication and much less likely with phaco-

emulsification. The source of the bleeding is a ruptured long or short posterior ciliary artery. Although the exact cause is unknown, contributing factors include advanced age, glaucoma, increased axial length, systemic cardiovascular disease and vitreous loss.

1. **Signs** (in chronological order)
   - Progressive shallowing of the anterior chamber, increased intraocular pressure and prolapse of the iris.
   - Vitreous extrusion, loss of the red reflex and the appearance of a dark mound behind the pupil.
   - In severe cases, all intraocular contents may be extruded through the incision.
2. **Immediate treatment** involves closure of the incision. Although posterior sclerotomy has been advocated, it may actually exacerbate the bleeding and result in a vicious circle with loss of the eye. Postoperatively, the patient should be treated with topical and systemic steroids to reduce intraocular inflammation.
3. **Subsequent management**
   a. ***Ultrasonography*** may be used to assess severity (Fig. 8.31).
   b. ***Surgery*** is performed 7–14 days later when the blood clot has liquefied. The blood is drained, and pars plana vitrectomy with air–fluid exchange performed (Fig. 8.32). Although the visual prognosis is grave, useful vision may be salvaged in some cases.

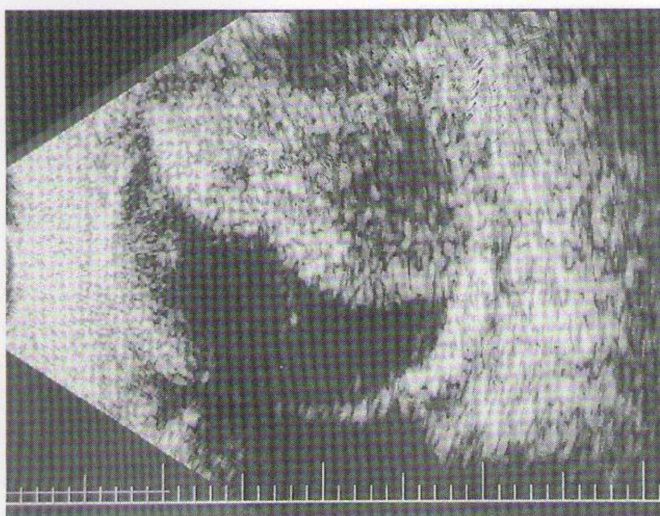


Fig. 8.31
Ultrasonogram showing a large suprachoroidal haemorrhage

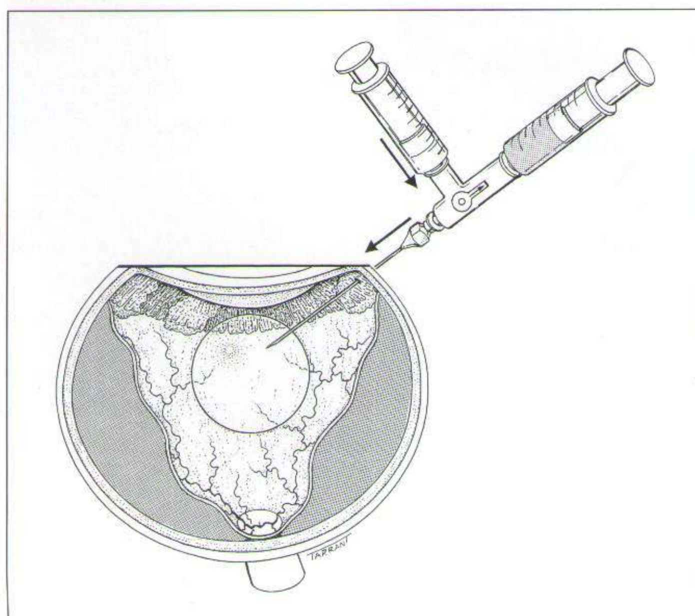


Fig. 8.32
Air–fluid exchange following vitrectomy for expulsive haemorrhage

Acute postoperative endophthalmitis

Causes

Acute endophthalmitis is a devastating complication that occurs in about 1:1000 cases.

1. **Causative organisms**, in order of frequency, include coagulase-negative staphylococci (e.g. *Staph. epidermidis*), other Gram-positive organisms (e.g. *Staph. aureus*) and Gram-negative organisms (e.g. *Pseudomonas* sp., *Proteus* sp.).
2. **The source of infection** most often cannot be identified with certainty. It is thought that the patient's own external bacterial flora of the eyelids, conjunctiva and lacrimal drainage passages is the most frequent culprit. Other potential sources of infection include contaminated solutions and instruments, and environmental flora including that of the surgeon and operating room personnel.

Prevention

Despite numerous studies, optimum prophylaxis has not yet been determined. The following measures may be beneficial:

1. **Preoperative treatment** of pre-existing infections such as staphylococcal blepharitis, conjunctivitis, dacryocystitis and infected contralateral sockets in patients with ocular prostheses (Fig. 8.33).
2. **Povidone–iodine** instilled preoperatively as follows:
   a. A 5% solution is prepared by diluting commercially available 10% Betadine aqueous solution used for skin



Fig. 8.33
Infected left socket of an artificial eye

preparation with an equal volume of balanced salt solution.

b. Two drops of the diluted solution are instilled into the conjunctival sac several minutes prior to surgery (Fig. 8.34) and the eyelids gently manipulated to distribute the solution over the ocular surface. The solution is also used to paint the skin of the eyelids, prior to draping (Fig. 8.35).

c. Prior to commencing surgery, the eye is irrigated with saline solution.

3. **Meticulous draping technique** that ensures that the lashes and lid margins are isolated (Fig. 8.36).

4. **Prophylactic antibiotics**
   a. *Postoperative injection* of anterior sub-Tenon antibiotics is commonly performed, but evidence of its efficacy is scant.
   b. *Intraoperative irrigation* of the anterior chamber by adding antibiotics such as vancomycin into the infusion fluid may be efficacious but may also lead to the emergence of resistant strains of bacteria.



Fig. 8.34
Preoperative instillation of povidone–iodine

Clinical features

The severity of endophthalmitis reflects the virulence of the offending organism.

1. **Extremely** severe endophthalmitis is characterized by pain, marked visual loss, lid oedema, chemosis,

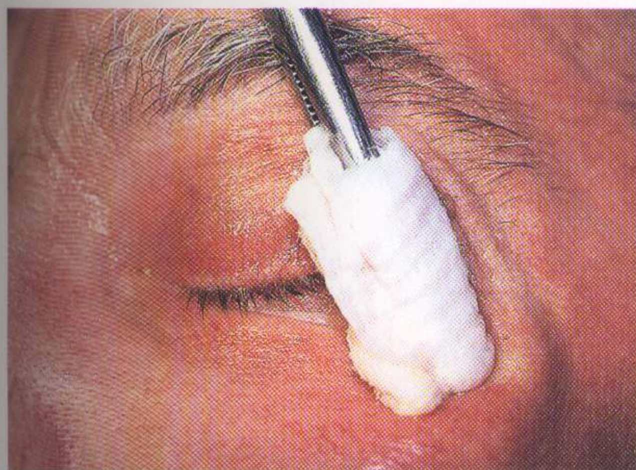


Fig. 8.35
Preparation of the skin with povidone–iodine



Fig. 8.37
Large hypopyon in acute postoperative endophthalmitis

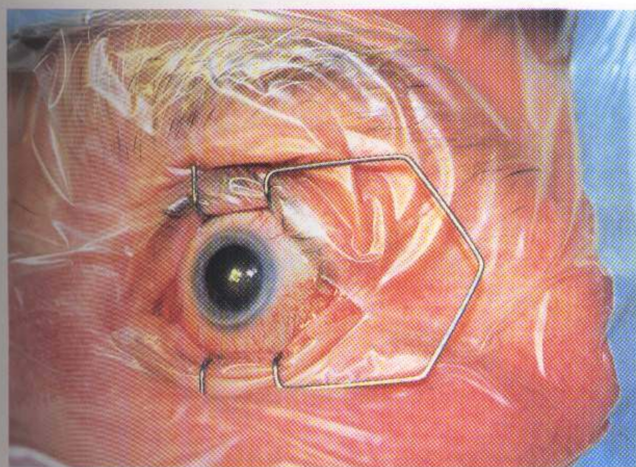


Fig. 8.36
Drapes isolating the eyelids from the operating field

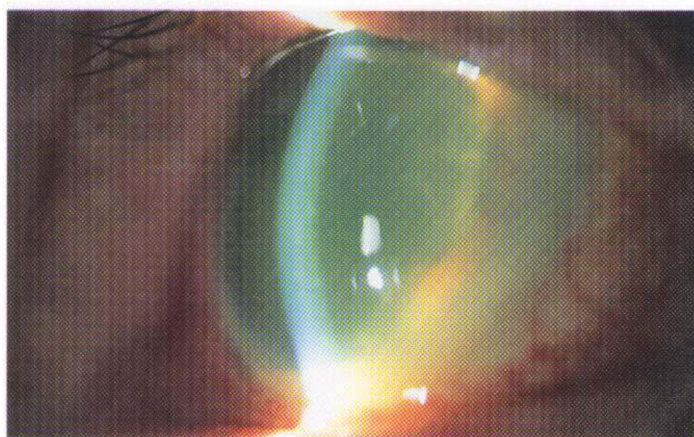


Fig. 8.38
Fibrinous exudate and small hypopyon in acute postoperative endophthalmitis

conjunctival injection, purulent discharge, corneal infiltrates and a large hypopyon (Fig. 8.37).

2. **Moderately** severe endophthalmitis is characterized by fibrinous exudate in the anterior chamber, a smaller hypopyon (Fig. 8.38), vitritis, absent red reflex and inability to visualize the fundus even with the indirect ophthalmoscope (Fig. 8.39).
3. **Mild** endophthalmitis may be associated with only slight pain, absent or small hypopyon (Fig. 8.40) and preservation of some red reflex with ability to visualize some fundus details with the indirect ophthalmoscope.

> *NB:* The time interval between surgery and onset of symptoms may be useful in predicting the likely offending organisms. For example:

- *Staph. aureus* and Gram-negative bacilli typically present 2–4 days postoperatively with florid endophthalmitis.
- *Staph. epidermidis* and coagulase-negative cocci usually present 5–7 days postoperatively with relatively milder signs.

Differential diagnosis

1. **Retained lens material** in the anterior chamber (Fig. 8.41) or vitreous may be associated with a severe anterior uveitis.
2. **Toxic reaction** to irrigating fluid or foreign material introduced into the eye at the time of surgery may occur. Rarely, an intense fibrinous reaction may develop on the anterior surface of the IOL (Fig. 8.42). Intensive topical and periocular steroids combined with cycloplegics are effective, although synechiae to the IOL may develop.
3. **Difficult or prolonged surgery** may result in corneal oedema and uveitis. This however, is evident in the immediate postoperative period.

Management

1. **Identification of causative organisms** from aqueous and vitreous confirms the diagnosis. However, negative culture does not necessarily rule out infection. The samples should be taken in the operating room as follows:

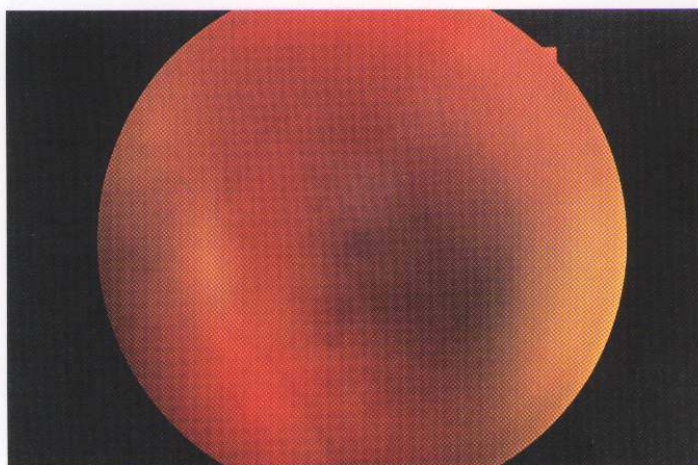


Fig. 8.39
Vitreous haze in acute postoperative endophthalmitis

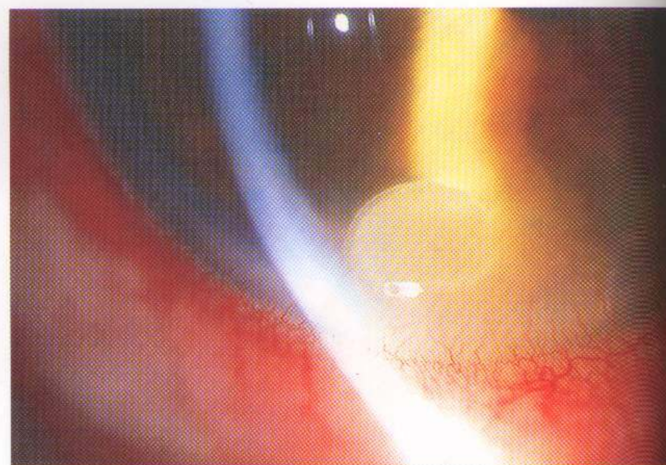


Fig. 8.41
Retained lens material in the anterior chamber

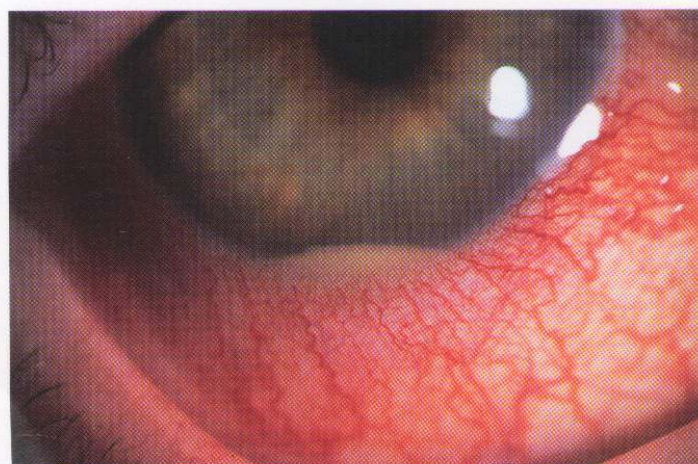


Fig. 8.40
Small hypopyon in acute postoperative endophthalmitis

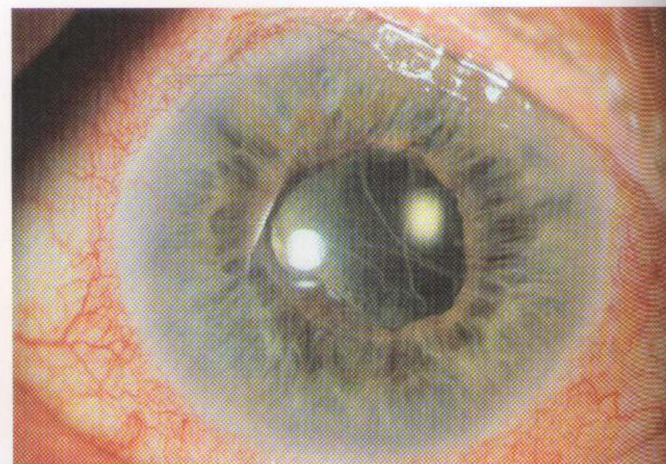


Fig. 8.42
Sterile fibrinous reaction following cataract extraction

a. *Aqueous samples* are obtained by aspirating 0.1 ml of aqueous with a needle on a tuberculin syringe, through the pre-existing 'second incision'.

b. *Vitreous samples* are ideally obtained with a mini-vitrector (Fig. 8.43) through a 'pars plana' sclerotomy 3.5 mm behind the limbus (Figs 8.44 and 8.45). If a mini-vitrector is unavailable, an alternative method involves a partial-thickness sclerotomy 3.5 mm behind the limbus, with aspiration of liquid vitreous from the mid-vitreous cavity with a needle attached to a tuberculin syringe. Between 0.1 and 0.3 ml of vitreous is removed and inoculated onto blood agar, chocolate agar, liquid thioglycolate and Sabouraud agar. If these culture media are not readily available, direct inoculation of blood culture bottles is a good alternative. Several drops are also placed onto slides for Gram and Giemsa staining.

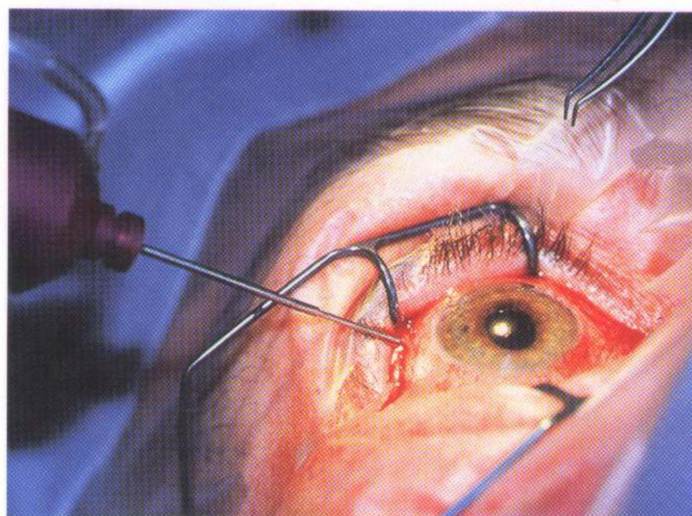


Fig. 8.45
Mini-vitrector obtaining vitreous samples

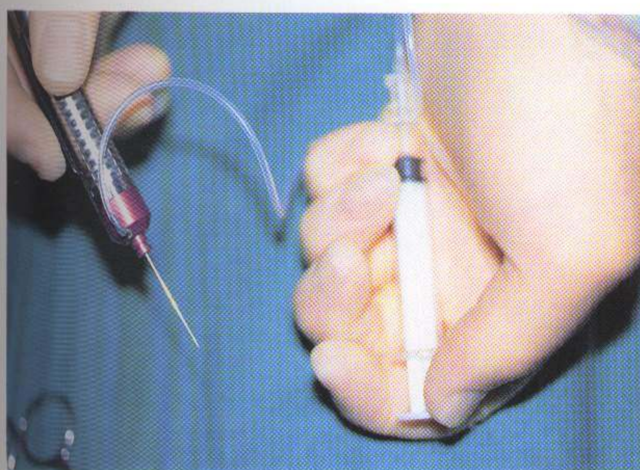


Fig. 8.43
Mini-vitrector for obtaining vitreous samples

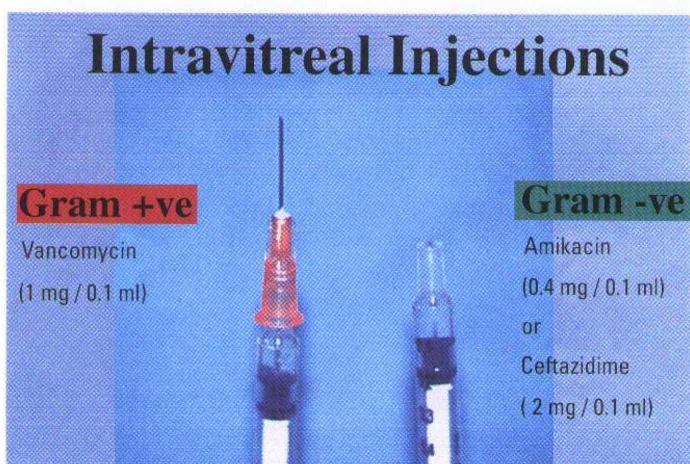


Fig. 8.46
Syringes containing antibiotics for intravitreal injection

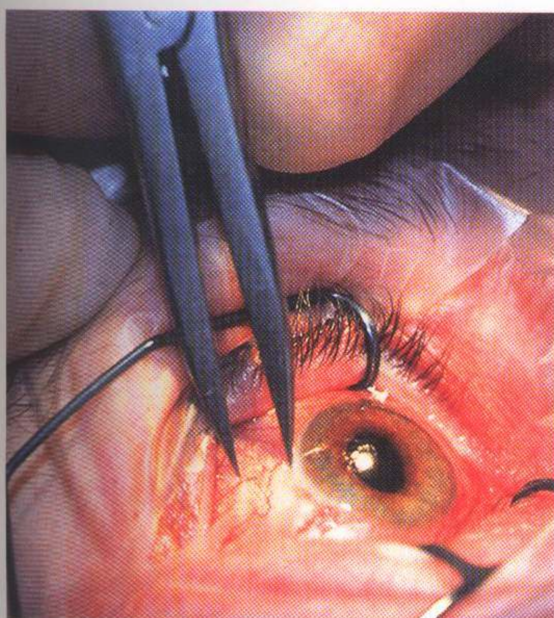


Fig. 8.44
Calipers measuring 3.5 mm from the limbus

2. **Vitrectomy** is beneficial only in the context of severe infection with visual acuity reduced to 'light perception'. If visual acuity is 'hand movements' or better then vitrectomy is unnecessary.
3. **Antibiotics** used are amikacin or ceftazidime, to cover most Gram-positive and Gram-negative organisms, and vancomycin for coagulase-negative and coagulase-positive cocci (Fig. 8.46). Amikacin acts synergistically with vancomycin but is potentially more retinotoxic than ceftazidime, which is however, not synergistic with vancomycin.
   a. *Intravitreal* antibiotics should be administered immediately after culture specimens have been obtained and the eye has been softened. Amikacin (0.4 mg in 0.1 ml) or ceftazidime (2.0 mg in 0.1 ml) and vancomycin (1 mg in 0.1 ml) are injected slowly into the mid-vitreous cavity using a 25-gauge needle (Fig. 8.47). The bevel of the needle should face anteriorly so as to minimize contact between the drug and the macula. After the first injection has been given, the syringe is disconnected but the needle is left inside the vitreous cavity so that the second injection can be given through the same needle. If

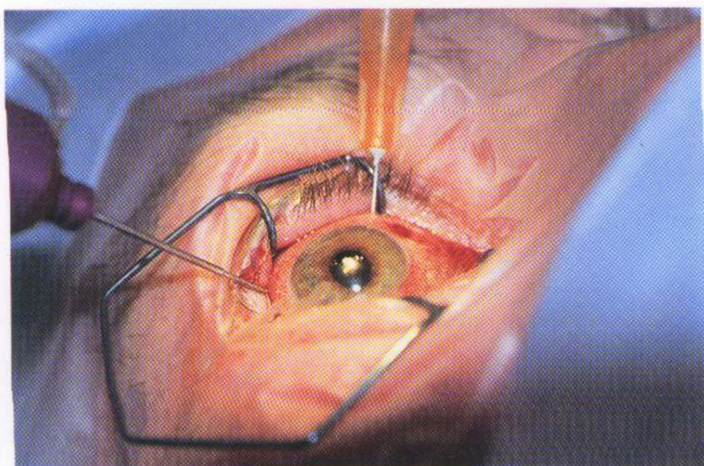


Fig. 8.47
Intravitreal injection of antibiotics



Fig. 8.48
Contraction of exudate following successful treatment

there is concern about the possibility of the formation of precipitates then two separate needles, each with a different antibiotic, may be used. The needle is then removed and a periocular injection of antibiotic given.

b. ***Periocular*** injections of vancomycin 25 mg and ceftazidime 100 mg, or gentamicin 20 mg and cefuroxime 125 mg, result in therapeutic intraocular concentrations and should be used. The injections are repeated daily for 5–7 days according to the response to therapy.

c. ***Topical*** therapy is of limited benefit in the absence of associated infectious keratitis.

d. ***Systemic*** therapy is controversial. The Endophthalmitis Vitrectomy Study Group showed that systemic ceftazidime and amikacin were not beneficial. These water-soluble antibiotics also have relatively poor coverage for Gram-positive organisms and poor ocular penetration. It is therefore possible that other antibiotics, such as the lipid-soluble quinolones (e.g. ciprofloxacin, ofloxacin) and imipenem, which have better penetration and a larger antimicrobial spectrum, may be beneficial. This can only be answered by future studies.

4. **Steroid** therapy should be commenced after antibiotics have been administered to limit the amount of inflammatory-induced damage. Steroids will not interfere with the control of the infection, provided the organisms are sensitive to the antibiotics.
   a. ***Periocular*** betamethasone 4 mg or dexamethasone 4 mg (1 ml) is administered daily for 5–7 days according to response to therapy.
   b. ***Oral*** prednisolone 20 mg q.i.d. for 10–14 days may be considered in very severe cases.
   c. ***Topical*** 0.1% dexamethasone is initially given every 30 minutes and then less frequently.
5. **Subsequent management** is to a certain extent governed by culture results and clinical findings.
   - Signs of improvement include reduction of anterior chamber cellular activity and hypopyon, and contraction of fibrinous exudate (Fig. 8.48). In this situation treatment is not modified irrespective of culture results.
   - If resistant bacteria are cultured and the clinical findings are worsening, antibiotic therapy should be modified accordingly. However, it is often too late.
6. **Results** of treatment are poor despite aggressive and appropriate therapy, with 55% of eyes achieving a final visual acuity of 6/60 or less.

NB: In some cases poor vision may be caused by antibiotic retinotoxicity, particularly aminoglycosides. Figure 8.49 shows macular infarction following intravitreal antibiotic injection. Figure 8.49a shows pallor of the posterior pole with haemorrhages. The fluorescein angiogram (Figs 8.49b–d) shows hypofluorescence due to ischaemia.

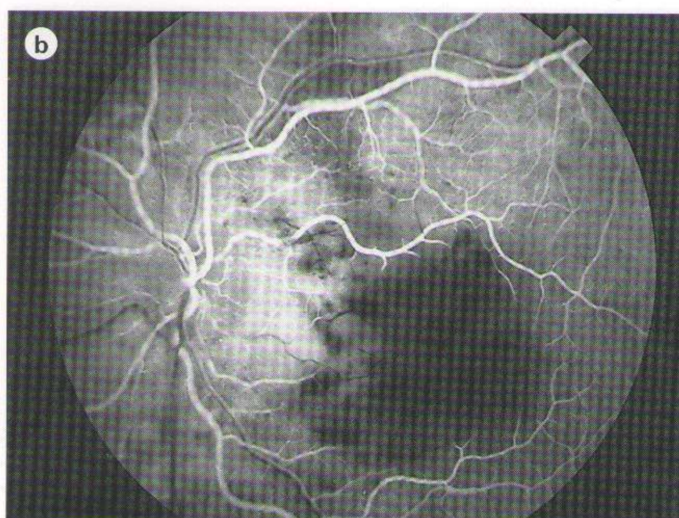
Delayed chronic postoperative endophthalmitis

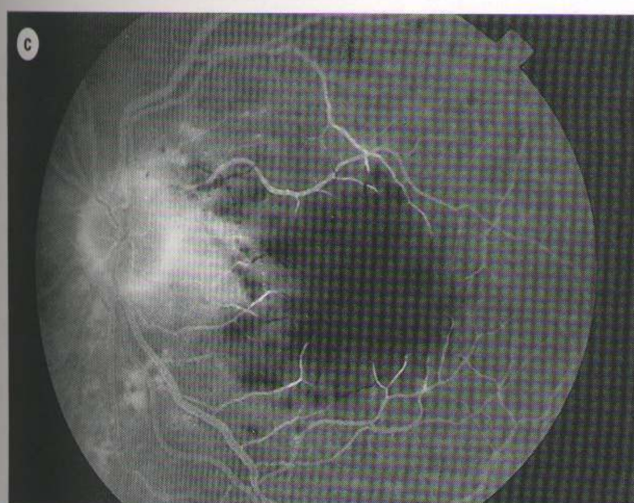
Delayed chronic indolent endophthalmitis occurs when an organism of low virulence becomes trapped within the capsular bag. It has an onset ranging from 4 weeks to years (mean of 9 months) postoperatively and typically follows uneventful cataract extraction with a PC-IOL. It may rarely be precipitated by YAG laser capsulotomy, which releases the sequestrated organisms from the posterior capsule into the vitreous. The infection is caused most frequently by *Propionibacterium acnes* and occasionally *Staph. epidermidis*, *Actinomyces israelii* and *Corynebacterium* spp.

Clinical features

1. **Presentation** is with mild progressive visual deterioration which may be associated with floaters but without pain.
2. **Signs**
   - Low-grade anterior uveitis, sometimes with granulomatous features such as mutton-fat keratic precipitates (Fig. 8.50).
   - Vitritis is common but hypopyon occurs infrequently.







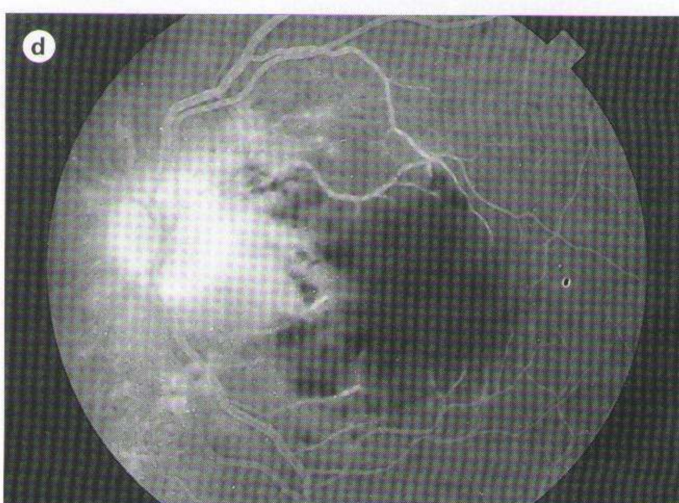


Fig. 8.49
Macular ischaemia following intravitreal antibiotic injection (*see text*)



Fig. 8.50
Mutton-fat keratic precipitates in chronic postoperative endophthalmitis

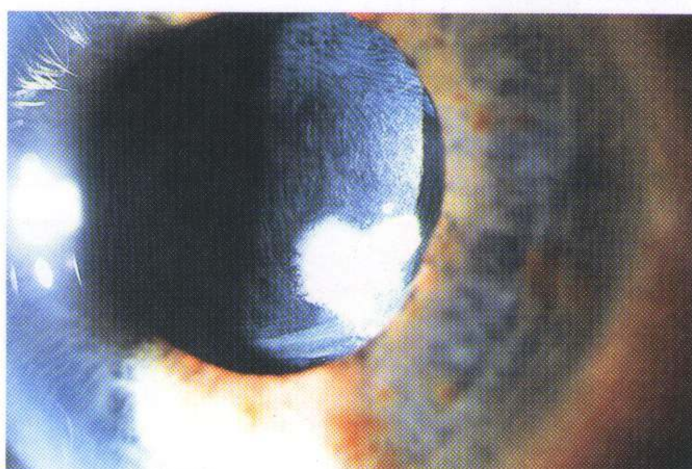


Fig. 8.51
White capsular plaque in chronic postoperative endophthalmitis

- An enlarging white capsular plaque is highly suggestive of infection by *Propionibacterium acnes*. It is composed of sequestered collections of organisms inside the peripheral capsular bag (Fig. 8.51).

> ***NB:*** It is important to perform gonioscopy under mydriasis so as not to miss an equatorial plaque.



Fig. 8.52
Fewer keratic precipitates following topical steroid therapy in chronic postoperative endophthalmitis

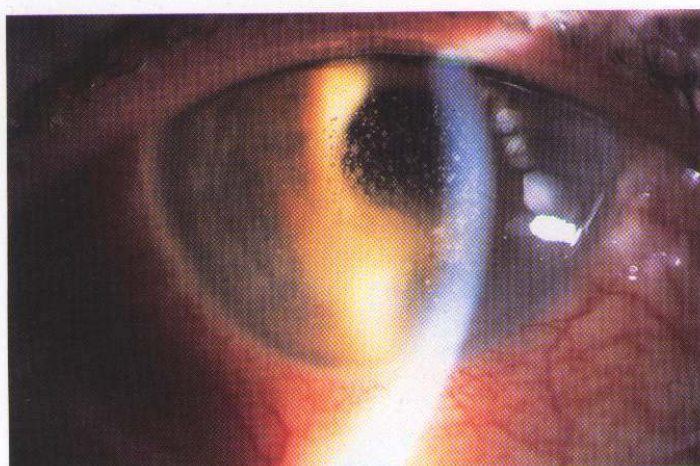


Fig. 8.53
Severe recurrence of inflammation 2 weeks following cessation of steroid therapy

3. **Clinical course**. The inflammation initially responds well to topical steroids (Fig. 8.52), but recurs after cessation of treatment (Fig. 8.53) and then becomes refractory.

Investigations

The diagnosis should be confirmed by cultures of the aqueous and vitreous with growth of the organisms on thioglycolate broth. Unfortunately this frequently fails to show the causative organism, mainly because of the small number and low pathogenicity of the pathogens, which may take 10–14 days to grow. Detection of pathogens can be greatly improved with the polymerase chain reaction (PCR).

Treatment

This is difficult because the sequestered organisms are isolated from host defences and antibiotics.

1. Topical and periocular steroids, and antibiotics may be tried but the response is transient.
2. Intravitreal vancomycin (1 mg in 0.1 ml) alone or combined with pars plana vitrectomy is successful in 50%.
3. Removal of the capsular bag, residual cortex and IOL may eventually be required. Secondary IOL implantation may be considered at a later date.

Postoperative capsular opacification

Posterior capsular opacification

Visually significant posterior capsular opacification (PCO) is the most common late complication of uncomplicated cataract surgery. Apart from reducing visual acuity, PCO may impair contrast sensitivity, cause difficulties with glare or give rise to monocular diplopia. Certain acrylic IOLs may be associated with lower rates of PCO than PMMA and silicone lenses. Implant design may also be relevant in this context: notably, a square 'edge' to the optic appears to inhibit PCO.

1. **Clinical features**
   a. ***Elschnig pearls*** (bladder cells, Wedl cells) are caused by the proliferation and migration of residual equatorial epithelial cells along the posterior capsule at the site of apposition between the remnants of the anterior capsule and the posterior capsule. They impart a vacuolated appearance to the posterior capsule, best visualized on retroillumination (Fig. 8.54). This is the most frequently seen type of opacification and is related to the patient's age. It is extremely common in children if a posterior capsulorhexis is not performed at the time of surgery.
   b. ***Capsular fibrosis*** (Fig. 8.55), due to fibrous metaplasia of epithelial cells, is less common and usually appears earlier than Elschnig pearls.
2. **Treatment** involves the creation of an opening in the posterior capsule with the Nd:YAG laser. Indications for capsulotomy include:
   - Diminished visual acuity.
   - Diplopia or glare, secondary to capsular wrinkling.
   - Inadequate fundus view impairing diagnosis, monitoring or treatment of retinal pathology.

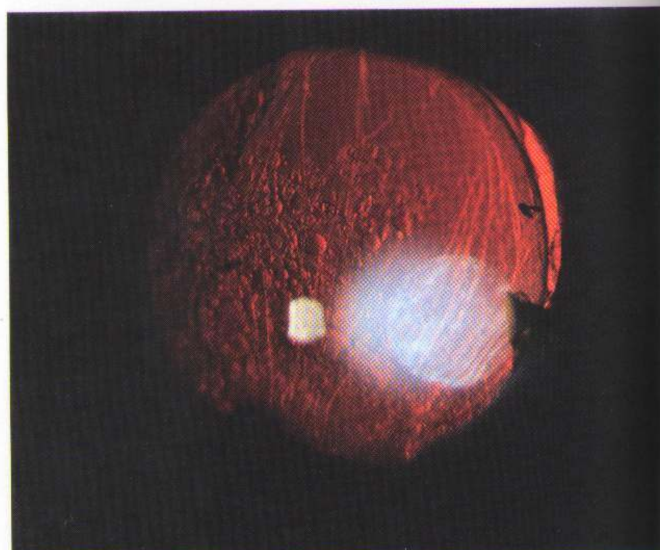


Fig. 8.54
Elschnig pearls

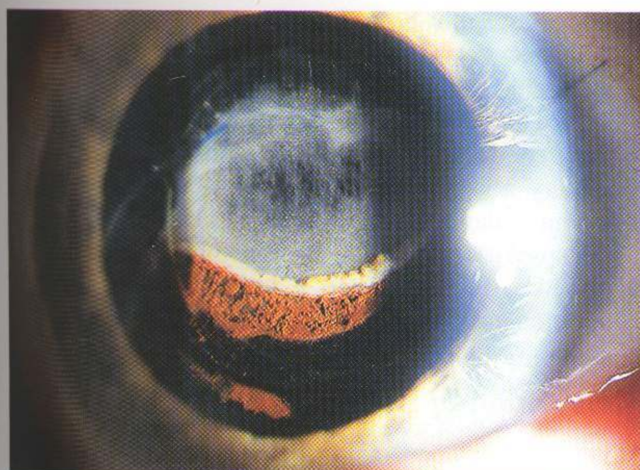


Fig. 8.55
Capsular fibrosis

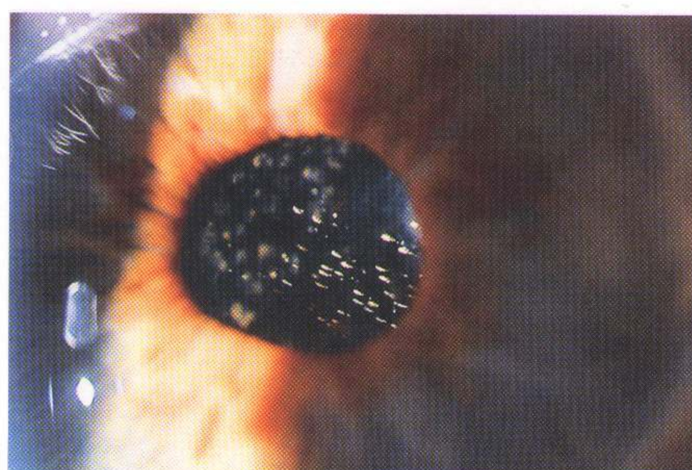


Fig. 8.57
Laser pitting of the intraocular implant

3. **Technique**. Safe and successful laser capsulotomy involves accurate focusing and using the minimum energy required. Laser power is initially set at 1 mJ/pulse, and may be increased if necessary. A series of punctures are applied in a cruciate pattern, the first puncture aimed at the visual axis (Fig. 8.56). An opening of about 3 mm is usually adequate, but larger capsulotomies may be necessary for retinal examination or photocoagulation.
4. **Complications**
   a. ***Damage to the IOL*** ('pitting') (Fig. 8.57) may occur if the laser is poorly focused. Although undesirable, a few laser marks on the IOL do not alter visual function or impair ocular tolerance of the IOL.
   b. ***Cystoid macular oedema*** is an occasional complication and may develop months after capsulotomy. It is less common when capsulotomy is delayed for 6 months or more after cataract surgery.
   c. ***Rhegmatogenous retinal detachment*** is rare, except in high myopes, and may occur several months after capsulotomy.
   d. ***Intraocular pressure elevation***, which is mild and transient, is usually innocuous. However, sustained elevation above precapsulotomy levels may occur, especially in patients with established glaucoma or those that manifest significant ocular hypertension within hours of the capsulotomy.
   e. ***Posterior IOL subluxation or dislocation*** (Fig. 8.58) is rare but may occur, particularly with plate haptic silicone and hydrogel IOLs.
   f. ***Chronic endophthalmitis*** due to release of sequestered organisms into the vitreous is very rare (*see* above).

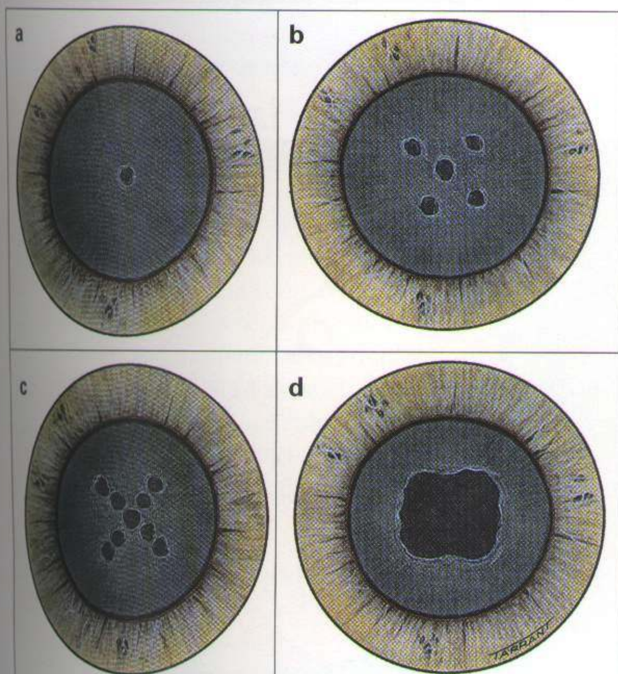


Fig. 8.56
Technique of Nd:YAG laser posterior capsulotomy

> ***NB:*** Most patients who develop complications have no identifiable risk factor. The number of laser pulses and the energy level are probably not related to their development.



Fig. 8.58
Dislocation of an intraocular implant



Fig. 8.59
Anterior capsular opacification and capsulophimosis

Anterior capsular opacification

Anterior capsular opacification (ACO), also called anterior capsular fibrosis or anterior subcapsular opacification, is less common but occurs earlier than PCO, sometimes within 1 month postoperatively. Severe fibrosis may result in anterior capsular shrinkage and constriction of the capsulorhexis (capsulophimosis) (Fig. 8.59). IOL design and material may be relevant in the development of ACO. The highest rate is with plate-haptic silicone IOLs and the lowest with three-piece acrylic optic–PMMA haptic IOLs. A small capsulorhexis may also be of relevance.

Miscellaneous postoperative complications

Corneal oedema

Corneal oedema (*see* Fig. 5.9) is usually transient and often caused by intraoperative trauma to the endothelium by contact with lens matter, instruments or IOL. Eyes with pre-existing Fuchs endothelial dystrophy are at increased risk. Other causes include excessive power use during phacoemulsification, complicated or prolonged surgery, and postoperative ocular hypertension.

Iris prolapse

Iris prolapse is extremely rare following small incision surgery but may occur after ECCE (Fig. 8.60).

1. **Causes**
   - A phaco incision with a peripheral point of entry into the anterior chamber.
   - A leaking incision.
   - Inadequate suturing of an ECCE incision.
   - Patient-related factors such as coughing and straining.
2. **Clinical features**
   - Prolapsed uveal tissue may be visible on the ocular surface.
   - The anterior chamber may be shallow in the area of the incision.
   - The pupil may be peaked (drawn) towards the prolapse.
3. **Complications** include defective wound healing, excessive astigmatism, epithelial ingrowth, chronic anterior uveitis, cystoid macular oedema and endophthalmitis.
4. **Treatment** depends on the time interval between cataract surgery and identification of the prolapse. If discovered within a day or two, with no evidence of infection, the prolapsed iris may be reposited and the incision resutured. If, however, the prolapse is of longer duration, the risk of infection mandates that the prolapsed iris be excised.

Malposition of IOL

Although uncommon, malposition may be associated with both optical and structural problems. Annoying visual aberrations include glare, haloes, and monocular diplopia if the edge of the IOL becomes displaced into the pupil (Fig. 8.61).

1. **Causes**
   - Primary malposition occurs during initial surgery. This may be due to zonulodialysis, capsular rupture, or after

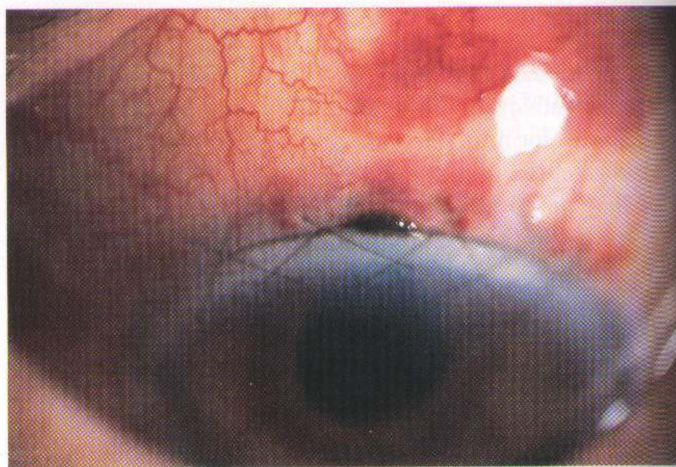


Fig. 8.60
Iris prolapse

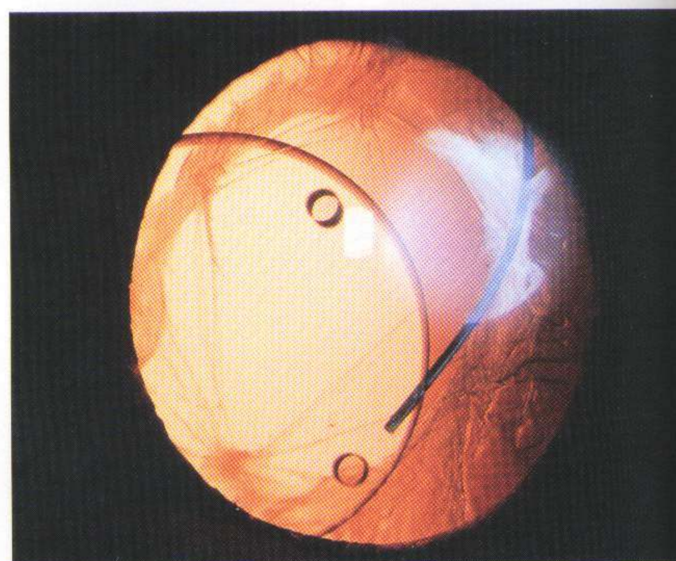


Fig. 8.61
Decentration of an intraocular implant

routine phacoemulsification, when one haptic is inserted into the capsular bag and the other into the ciliary sulcus.

- Postoperative causes include trauma, eye rubbing and capsular contraction.

2. **Treatment** with miotics may be successful in mild cases. Significant malposition may necessitate replacement of the IOL.

Retinal detachment

Rhegmatogenous retinal detachment, although uncommon following uneventful ECCE or phaco, may be associated with the following risk factors:

1. **Preoperative**
   - Lattice degeneration or retinal breaks should be treated prophylactically prior to cataract surgery or laser capsulotomy if fundus view permits, or as soon as possible thereafter.
   - High myopia.
2. **Operative**
   - Disruption of the posterior capsule.
   - Vitreous loss, particularly if managed inappropriately, is associated with an approximate 7% risk of retinal detachment. Myopia of over 6D increases the risk to 15%.
3. **Postoperative**. YAG laser capsulotomy, if performed within a year of cataract surgery.

Cystoid macular oedema

This occurs commonly after complicated surgery involving rupture of the posterior capsule and vitreous prolapse, sometimes with incarceration in the incision, although it may occur after uneventful surgery. It usually presents 2–6 months after surgery. The clinical features and management are discussed in Chapter 13.

Congenital cataract

Congenital cataracts occur in about 3:10,000 live births; two-thirds of cases are bilateral. The cause of cataract formation can be identified in about half of those with bilateral opacities. The most common cause is genetic mutation, usually AD. Other causes include chromosomal abnormalities such as Down syndrome, metabolic disorders such as galactosaemia and intrauterine insults such as rubella infection. Congenital cataract may also occur as part of a complex developmental disorder of the eye such as aniridia.

Cataract without systemic associations

Isolated hereditary cataracts

These account for about 25% of cases. The mode of inheritance is most frequently AD but may be autosomal recessive (AR) or X-linked (X-L). The morphology of the opacities and also frequently the need for surgery are usually similar in parent and offspring. About 10 loci for AD cataract have been mapped. Isolated inherited congenital cataracts carry a better visual prognosis than those with coexisting ocular and systemic abnormalities. This is because they are frequently partial at birth so that surgery may be delayed until the child is older, when there is a lower incidence of surgical complications and refractive correction is easier. Morphological classification of hereditary cataracts is based on the location of the opacities within the lens as follows:

1. **Zonular cataract** in which the opacity occupies a discrete zone in the lens may be:
   a. ***Nuclear***, in which the opacities are confined to the embryonic or fetal nuclei of the lens. Some patients have dense central opacities (Fig. 8.62) while others have fine pulverulent (dust-like) opacities (Fig. 8.63).
   b. ***Lamellar***, in which the opacity is sandwiched between clear nucleus and cortex. It may be associated with radial extensions known as riders (Fig. 8.64).
   c. ***Capsular***, in which the opacity is confined to the anterior or posterior capsule.



Fig. 8.62
Congenital nuclear cataract



Fig. 8.63
Congenital nuclear pulverulent cataract



Fig. 8.64
Congenital lamellar cataract

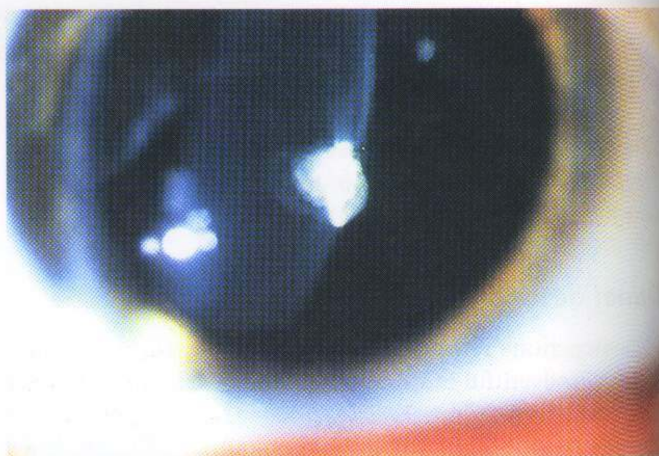


Fig. 8.67
Congenital anterior pyramidal cataract (Courtesy of A. Shun-Shin)

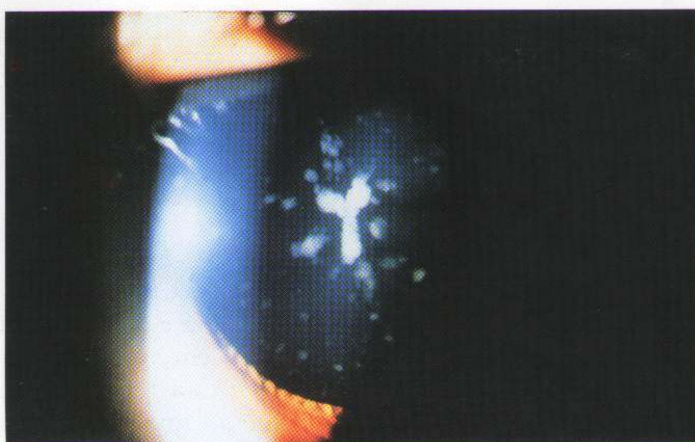


Fig. 8.65
Congenital sutural cataract



Fig. 8.68
Congenital anterior polar cataract and persistent pupillary membrane

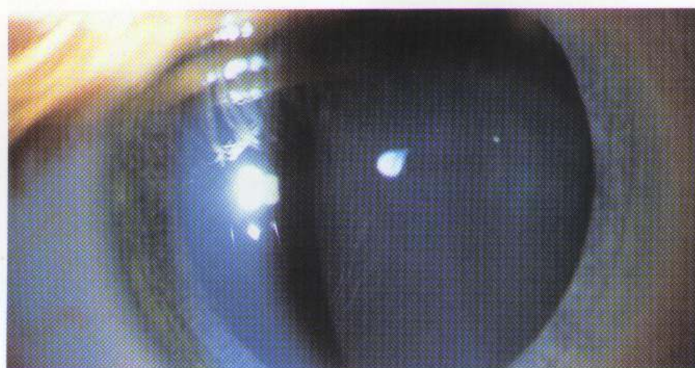


Fig. 8.66
Congenital flat anterior polar cataract



Fig. 8.69
Congenital posterior polar cataract

d. Sutural, in which the opacity follows the anterior or posterior Y suture. It may occur in isolation or in association with other opacities (Fig. 8.65).

2. **Polar cataract**, in which the opacities occupy the subcapsular cortex at the anterior or posterior pole of the lens.

a. Anterior polar cataract may be flat (Fig. 8.66) or project as a conical opacity into the anterior chamber (pyramidal cataract) (Fig. 8.67). Patients with pyramidal cataracts are likely to develop amblyopia due to either unilateral involvement or bilateral asymmetrical opacities. Occasional associations of

anterior polar cataracts include persistent pupillary membrane (Fig. 8.68), anterior lenticonus, Peters anomaly and aniridia.

b. Posterior polar cataract (Fig. 8.69) may occasionally be associated with persistent hyaloid remnants (Mittendorf dots), posterior lenticonus and persistent hyperplastic primary vitreous.

Other types

1. **Coronary (supranuclear)** cataract consists of round opacities in the deep cortex which surround the nucleus like a crown (Fig. 8.70). They are usually sporadic and only occasionally hereditary.
2. **Blue dot** opacities (cataracta punctata caerulea) (Fig. 8.71) are common and innocuous and may coexist with other types of lens opacities.
3. **Total** (mature) cataracts are frequently bilateral and often begin as lamellar or nuclear.
4. **Membranous** cataract (Fig. 8.72) is rare and may be associated with Hallermann–Streiff–Francois syndrome. It occurs when lenticular material partially or completely reabsorbs, leaving behind residual chalky-white lens matter sandwiched between the anterior and posterior capsules.

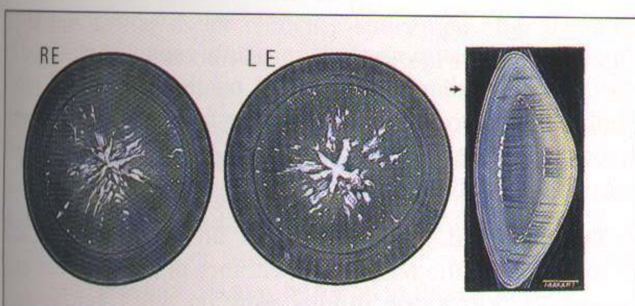


Fig. 8.70
Congenital coronary (supranuclear) cataracts

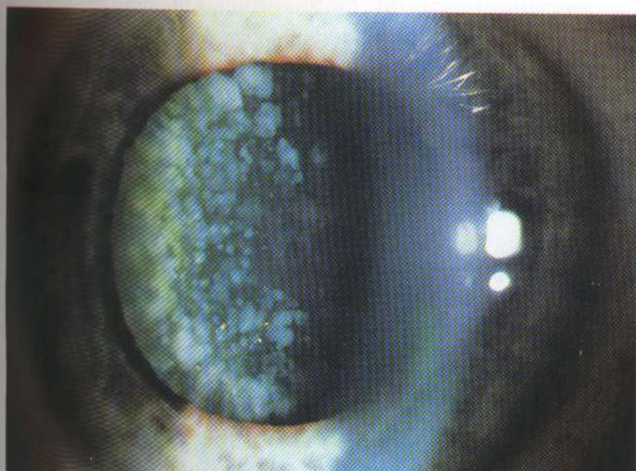


Fig. 8.71
Congenital blue dot cataract

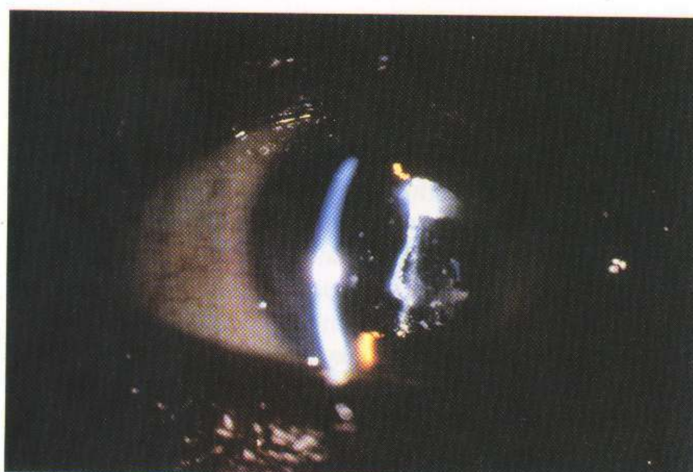


Fig. 8.72
Congenital membranous cataract

Systemic associations

A great many systemic paediatric conditions may be associated with congenital cataracts. The vast majority are extremely rare and of interest only to paediatric ophthalmologists. The general ophthalmologist should, however, be aware of the following conditions:

Metabolic

1. **Galactosaemia** involves severe impairment of galactose utilization caused by absence of the enzyme galactose-1-phosphate uridyl transferase (GPUT). Inheritance is AR.
   a. Systemic features, which become manifest during infancy, include failure to thrive, lethargy, vomiting and diarrhoea. Reducing substance is found in the urine after drinking milk. Unless galactose, in the form of milk and milk products, is withheld from the diet, hepatosplenomegaly, renal disease, anaemia, deafness and mental handicap occur subsequently, with ultimate death.
   b. Cataract, characterized by a central 'oil droplet' opacity (Fig. 8.73), develops within the first few days or weeks

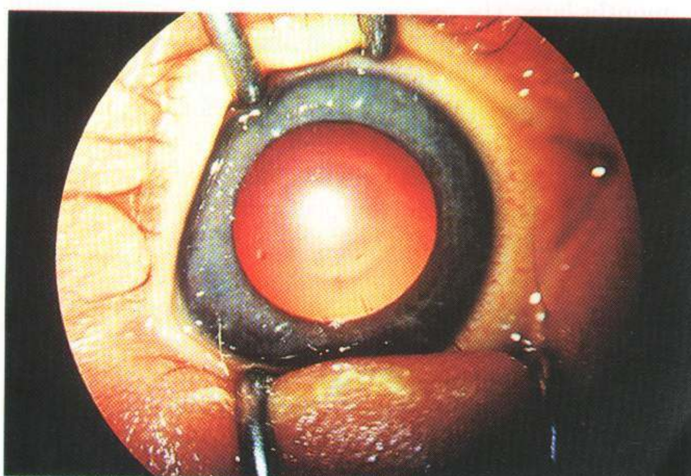


Fig. 8.73
'Oil droplet' cataract in galactosaemia (Courtesy of K. Nischal)

of life in a large percentage of patients. The exclusion of galactose (in milk products) from the diet will prevent the progression of cataract and may reverse early lens changes.

2. **Galactokinase deficiency** involves the first enzyme in the pathway of galactose metabolism. Inheritance is AR.
   a. *Systemic features* are absent although reducing substance is present in the urine after drinking milk.
   b. *Cataract*, consisting of lamellar opacities, may develop in the fetus or in early infancy. Some presenile cataracts may also result from galactokinase deficiency. Galactose is only indirectly cataractogenic as a result of its reduction to dulcitol within the lens. Dulcitol accumulation within the lens increases the intralenticular osmotic pressure with resultant osmotic influx of water, disruption of the lens fibres and opacification.
3. **Lowe (oculocerebrorenal) syndrome** is a rare inborn error of amino acid metabolism which predominantly affects boys. Inheritance is X-L.
   a. *Systemic features* include mental handicap, Fanconi syndrome of the proximal renal tubules, muscular hypotonia, frontal prominence and sunken eyes. It is one of the few conditions in which congenital cataract and congenital glaucoma may coexist.
   b. *Cataract* is universal; the lens is also small, thin and disc-like (microphakia) and may show posterior lentiglobus. Cataract may be capsular, lamellar, nuclear or total. Female carriers manifest micropunctate cortical lens opacities usually without visual impact.
   c. *Congenital glaucoma* is present in 50% of cases.
4. **Other** disorders include hypoparathyroidism, pseudohypoparathyroidism and mannosidosis.

Prenatal infections

1. **Congenital rubella** is associated with cataract in about 15% of cases. After the gestational age of 6 weeks, the virus is incapable of crossing the lens capsule so that the lens is immune. Although the lens opacities (which may be unilateral or bilateral) are usually present at birth, they may occasionally develop several weeks or even months later. The opacity may involve the nucleus, with a dense pearly appearance, or may present as a more diffuse opacity involving most of the lens. The virus is capable of persisting within the lens for up to 3 years after birth.
2. **Other** intrauterine infections that may be associated with neonatal cataract are toxoplasmosis, cytomegalovirus, herpes simplex and varicella.

Chromosomal abnormalities

1. **Down syndrome** (trisomy 21)
   a. *Systemic features* include mental handicap, upward-slanting palpebral fissures, epicanthic folds, flat midface with relative prognathism, brachycephalic skull with flattening of the occiput, broad and short hands and a protruding tongue.
   b. *Cataract* of various morphology occurs in about 5% of patients. The opacities are usually symmetrical and often develop late in childhood.
2. **Other** chromosomal abnormalities associated with cataract include Patau (trisomy 13) and Edward (trisomy 18) syndromes.

Skeletal syndromes

1. **Hallermann–Streiff–Francois syndrome** is a sporadic disorder.
   a. *Systemic features* include frontal prominence, small beaked nose, baldness, progeria, micrognathia and pointed chin, short stature and hypodontia.
   b. *Cataract*, which may be membranous (*see* Figure 8.72), occurs in 90% of cases.
2. **Nance–Horan syndrome** is an X-L disorder.
   a. *Systemic features* include supernumerary incisors, prominent ears, anteverted pinnae and shortened metacarpals.
   b. *Cataract* may be dense and associated with mild microphthalmos. Female carriers may show mild sutural lens opacities.

Management

Ocular examination

Since a formal estimate of visual acuity cannot be obtained in the neonate, reliance is required on the density and morphology of the opacity, other ocular associated findings and visual behaviour of the child, in order to assess the visual significance of the cataract.

1. **Density** and potential impact on visual function are assessed on the basis of appearance of the red reflex and the quality of the fundus view on direct and indirect ophthalmoscopy. However, slit-lamp examination of a neonate has been made easier with the introduction of high-quality portable slit-lamps. With assistance to restrain head movements, detailed anterior segment assessment should be possible. On ophthalmoscopy cataract density is graded as follows:
   - A very dense cataract occluding the pupil will preclude any view with either instrument and the decision to operate is straightforward.
   - A less dense cataract, which is, however, still visually significant, will allow visualization of the retinal vasculature with the indirect but not with the direct ophthalmoscope. Other features of visually significant cataract are central or posterior opacities over 3 mm in diameter.
   - A visually insignificant opacity will allow clear visualization of the retinal vasculature with both the indirect and direct ophthalmoscope. Other features of visually insignificant cataract are central opacities less than 3 mm in diameter and peripheral, anterior or punctate opacities with intervening clear zones.

2. **Morphology** of the opacity can give important clues to aetiology, as described previously.
3. **Associated ocular pathology** may involve the anterior segment (corneal clouding, microphthalmos, glaucoma, persistent hyperplastic primary vitreous) or the posterior segment (chorioretinitis, Leber amaurosis, rubella retinopathy, foveal or optic nerve hypoplasia). Examination under general anaesthesia may occasionally be required, as may repeated examinations, to document progression of cataract or associated disease.
4. **Other indicators** of severe visual impairment include absence of central fixation, nystagmus and strabismus. Nystagmus in particular signifies a poor visual prognosis.
5. **Special tests** such as forced choice preferential looking (*see* Chapter 16) and visually evoked potentials also provide helpful information but should not be relied upon exclusively since they may be misleading.

Systemic investigations

Unless there is an established hereditary basis for the cataracts, the investigation of the infant with bilateral cataracts should include the following:

1. **Serological tests** for intrauterine infections (TORCH = **to**xoplasmosis, **r**ubella, **c**ytomegalovirus and **h**erpes simplex). A history of maternal rash during pregnancy mandates the assay of varicella zoster antibody titres.
2. **Urine**. Urinalysis for reducing substance after drinking milk (galactosaemia) and chromatography for amino acids (Lowe syndrome).
3. **Other investigations** include fasting blood glucose, serum calcium and phosphorus, red blood cell GPUT and galactokinase levels.
4. **Referral to a paediatrician** may be warranted for dysmorphic features or suspicion of other systemic diseases. Chromosome analysis may be useful in this context.

Timing of surgery

This is crucial and the main considerations are as follows:

1. **Bilateral dense** cataracts (Fig. 8.74) require early surgery (i.e. by 6 weeks of age) to prevent the development of stimulus deprivation amblyopia. If the severity is asymmetrical, the eye with the denser cataract should be addressed first.



Fig. 8.74
Bilateral dense congenital cataracts

2. **Bilateral partial** cataracts may not require surgery until later, if at all. In cases of doubt it may be prudent to defer surgery, monitor lens opacities and visual function and intervene later if vision deteriorates.
3. **Unilateral dense** cataracts merit urgent surgery (within days) followed by aggressive anti-amblyopia therapy, despite which the results are often poor. If the cataract is detected after 16 weeks of age then surgery is inadvisable because amblyopia is refractory.
4. **Partial unilateral** cataracts can usually be observed or treated non-surgically with pupillary dilatation and possibly part-time contralateral occlusion to prevent amblyopia.

> ***NB:*** It is important to correct associated refractive errors.

Surgical technique

1. A 'scleral tunnel', 6 mm in width, is fashioned.
2. The anterior chamber is entered with a keratome (usually 3 mm wide) and the anterior chamber filled with viscoelastic substance.
3. An anterior capsulorhexis is performed. In children the anterior capsule is more elastic than in adults and the rhexis may be difficult due to a tendency to run outwards.
4. The lens matter is aspirated with a vitreous cutter or a Simcoe cannula.
5. A capsulorhexis is then performed on the posterior capsule.
6. A limited anterior vitrectomy is performed with a vitrector.
7. A PMMA posterior chamber IOL is implanted into the capsular bag, if appropriate. Some surgeons prolapse the optic through the posterior capsulotomy.
8. The viscoelastic substance is aspirated.
9. Intraocular scissors and forceps may be required to excise thick capsular material or retrolenticular plaques in eyes with associated persistent hyperplastic primary vitreous. Intraocular bipolar cautery may also be necessary to seal bleeding vessels.
10. A well-constructed scleral tunnel often will not require a suture.

Postoperative complications

Cataract surgery in children carries a higher incidence of complications than in adults.

1. **Posterior capsular opacification** is nearly universal if the posterior capsule is retained. It is also of more significance in young children because of its amblyogenic effect. Opacification of the anterior hyaloid face may occur despite posterior capsulorhexis if the anterior vitreous is left intact. The incidence of opacification is reduced when posterior capsulorhexis is combined with vitrectomy.

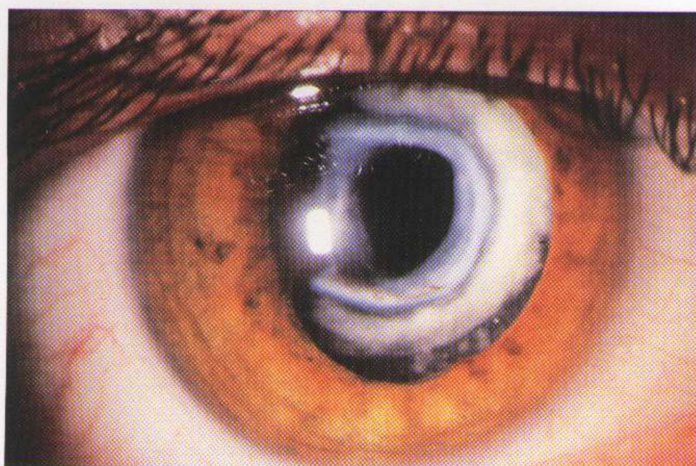


Fig. 8.75
Soemmerring ring following congenital cataract surgery

2. **Secondary membranes** may form across the pupil, particularly in microphthalmic eyes or those with associated chronic uveitis. A fibrinous postoperative uveitis in an otherwise normal eye, unless vigorously treated, may also result in membrane formation. Thin membranes may be opened with an Nd:YAG laser; thick ones may require surgery.
3. **Proliferation of lens epithelium** is universal but usually visually inconsequential, since it does not involve the visual axis. It becomes encapsulated within the remnants of the anterior and posterior capsules and is referred to as a Soemmerring ring (Fig. 8.75).
4. **Glaucoma** eventually develops in about 20% of eyes.
   - Closed-angle glaucoma may occur in the immediate postoperative period in microphthalmic eyes secondary to pupil block.
   - Secondary open-angle glaucoma may also develop years after the initial surgery. It is therefore important to monitor the intraocular pressure regularly for many years.
5. **Retinal detachment** is an uncommon and usually late complication.

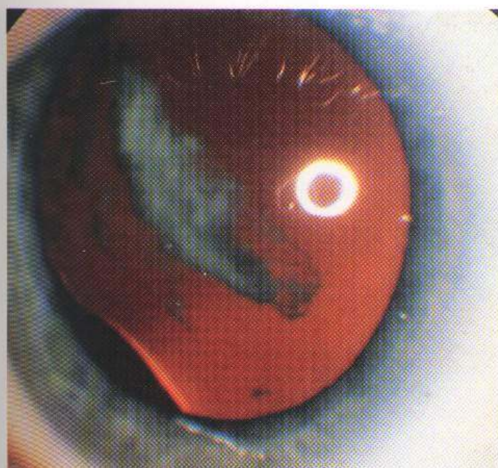
Visual rehabilitation

Although the technical difficulties of performing cataract surgery in infants and young children have mostly been resolved, visual results continue to be disappointing because of severe and irreversible amblyopia. With regard to optical correction for the aphakic child, the two main considerations are age and laterality of aphakia.

1. **Spectacles** are useful for older children with bilateral aphakia, but not for unilateral aphakia because of associated anisometropia and aniseikonia. Even in infants with bilateral aphakia they may be inappropriate because of their weight, unpleasant appearance, prismatic distortion and constriction of the visual field.
2. **Contact lenses** provide a superior optical solution for both unilateral and bilateral aphakia. Tolerance is usually reasonable until the age of about 2 years, although after this period problems with compliance may start as the child becomes more active and independent. The contact lens may become dislodged or lost, leading to periods of visual deprivation with the risk of amblyopia. In bilateral aphakia, the solution is simply to prescribe spectacles, although in unilateral aphakia IOL implantation may have to be considered.
3. **IOL implantation** is increasingly being performed in young children and even infants and appears to be effective and safe in selected cases. Awareness of the rate of myopic shift which occurs in the developing eye, combined with accurate biometry, allows the calculation of an IOL power targeted at initial hypermetropia (correctable with spectacles) which will ideally decay towards emmetropia later in life. However, final refraction is variable and emmetropia in adulthood cannot be guaranteed.
4. **Occlusion** to treat or prevent amblyopia is vital.

Anomalies of lens shape

1. **A lens coloboma** (Fig. 8.76) is characterized by notching (segmental agenesis) at the inferior equator. There is also a corresponding absence of zonular fibres. It is not a true coloboma as there is no focal absence of a tissue layer due to failure of closure of the optic fissure.
2. **Posterior lenticonus** (Fig. 8.77) is a very rare condition characterized by a round or conical bulge of the posterior axial zone of the lens into the vitreous, associated with local thinning or absence of the capsule. It may be associated with opacification of the posterior capsule and hyaloid remnants. With age, the bulge progressively increases in size and the lens cortex may opacify. Progression of cataract is variable, but many cases present with an acutely opacified white lens in infancy or early childhood. Most cases are unilateral, sporadic and not associated with systemic abnormalities. Rarely it may be bilateral and familial.
3. **Anterior lenticonus** (Fig. 8.78) is a bilateral axial projection of the anterior surface of the lens into the anterior chamber. About 90% of patients have Alport syndrome, which may also be associated with cataract, retinal flecks and posterior polymorphous corneal dystrophy.
4. **Lentiglobus** is a very rare, usually unilateral, generalized hemispherical deformity of the lens which may be associated with posterior polar lens opacity.
5. **Microphakia** (Fig. 8.79) is a lens with a smaller than normal diameter. It may be associated with Lowe syndrome, in which the lens is not only small but also disc-like.
6. **Microspherophakia** is a lens with a small diameter and spherical shape.
   a. Causes include familial (dominant) microspherophakia which is not associated with systemic defects, Marfan


Fig. 8.76
Coloboma of the lens

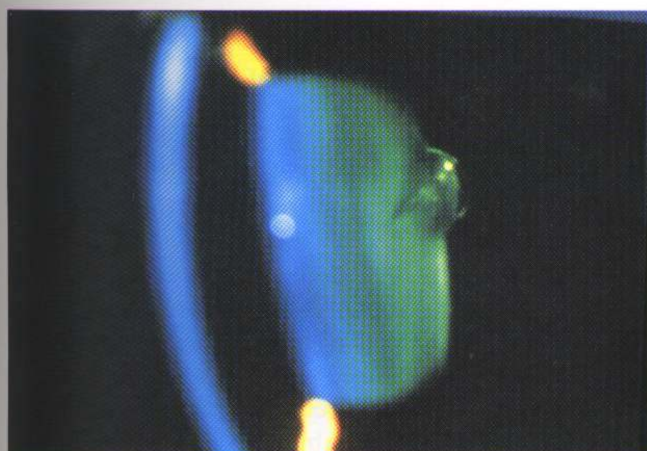
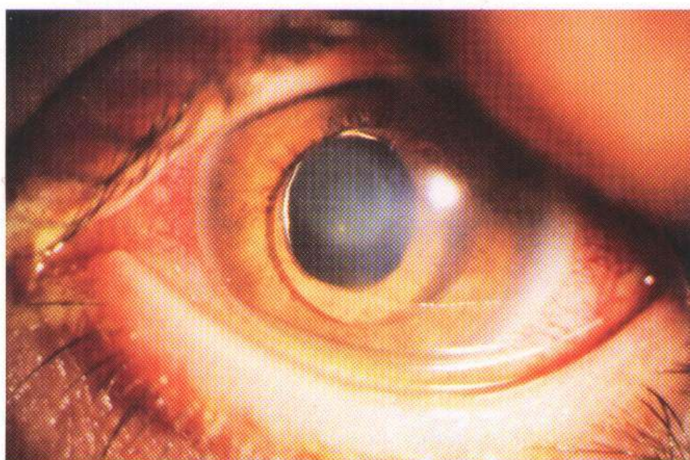

Fig. 8.77
Posterior lenticonus


Fig. 8.78
Anterior lenticonus (Courtesy of J. Govan)

syndrome, Weill–Marchesani syndrome, hyperlysinaemia and congenital rubella.

b. Ocular associations include Peters anomaly and familial ectopia lentis et pupillae.


Fig. 8.79
Microphakia


Fig. 8.80
Microspherophakic lens dislocated into the anterior chamber

c. Complications include lenticular myopia, subluxation and total dislocation into the anterior chamber (Fig. 8.80).

Ectopia lentis

Ectopia lentis refers to a displacement of the lens from its normal position. The lens may be completely dislocated, rendering the pupil aphakic (luxated), or partially displaced, still remaining in the pupillary area (subluxated). Ectopia lentis may be hereditary or acquired. Acquired causes include trauma, a large eye (i.e. high myopia, buphthalmos), anterior uveal tumours and hypermature cataract. Only hereditary causes are discussed below.

Without systemic associations

1. **Familial ectopia lentis** is characterized by bilateral symmetrical superotemporal lenticular displacement.

Inherited in an AD fashion, it may manifest congenitally or later in life.

2. **Ectopia lentis et pupillae** (Fig. 8.81) is a rare, congenital, bilateral, AR disorder characterized by displacement of the pupil and the lens in opposite directions. The pupils are small, slit-like and dilate poorly. Other findings include iris transillumination, enlarged corneal diameter, glaucoma, cataract and microspherophakia.
3. **Aniridia** may occasionally be associated with ectopia lentis.

With systemic associations

1. **Marfan syndrome** is a widespread, autosomal dominant, disorder of connective tissue (*see* Chapter 20) characterized by the following ocular features:
   a. *Ectopia lentis*, bilateral and symmetrical, is present in 80% of cases. Subluxation is most frequently superotemporal (Fig. 8.82), but may be in any meridian. Because the zonule is frequently intact, accommodation is retained, although rarely the lens may dislocate into the anterior chamber or vitreous (Fig. 8.83). The lens also may be microspherophakic.

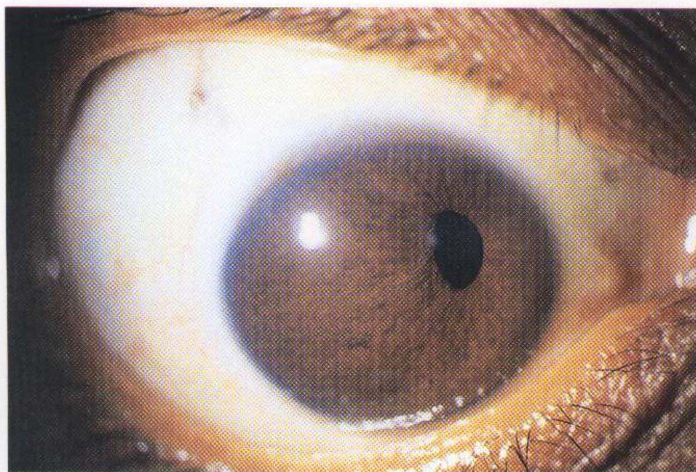


Fig. 8.81
Ectopia lentis et pupillae



Fig. 8.82
Upward lens subluxation in Marfan syndrome

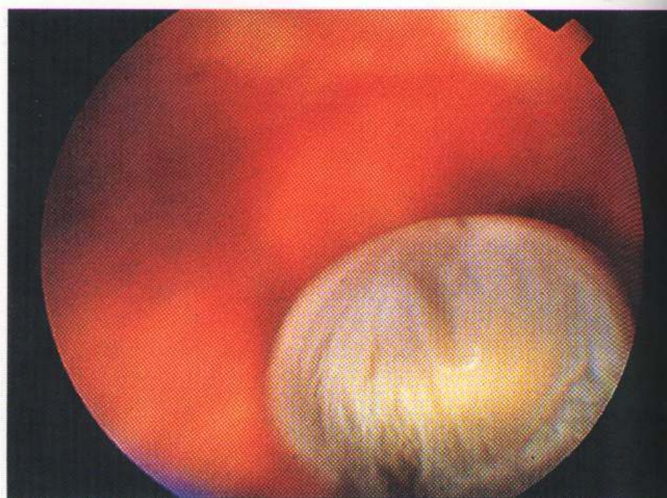


Fig. 8.83
Dislocation of the lens into the vitreous in Marfan syndrome
(Courtesy of S. Milewski)

   b. *Angle anomaly* is present in 75% of eyes. It is characterized by dense iris processes and thickened trabecular sheets, and may be responsible for glaucoma.
   c. *Retinal detachment* associated with lattice degeneration and high axial myopia is the most serious complication.
   d. *Other* features include hypoplasia of dilator pupillae (rendering mydriasis difficult), peripheral iris transillumination, strabismus, flat cornea and blue sclera.
2. **Weill–Marchesani syndrome** is a rare systemic connective tissue disease, conceptually the converse of Marfan syndrome, characterized by short stature, brachydactyly with stiff joints and mental handicap. Inheritance may be AD or AR.
   a. *Ectopia lentis*, bilateral and inferior, occurs in about 50% of cases during the 'teens' or early twenties. Microspherophakia is frequent.
   b. *Secondary angle-closure glaucoma* may occur due to pupillary block, consequent upon spherophakia and forward movement of the lens, although there may be a congenital angle anomaly.
   c. *Other* features include asymmetrical axial lengths and presenile vitreous liquefaction.
3. **Homocystinuria** is an AR inborn error of metabolism in which decreased hepatic activity of cystathionine β-synthetase results in systemic accumulation of homocysteine and methionine. Systemic features include skeletal anomalies with a Marfanoid habitus, fair hair and a tendency to thrombotic episodes.
   a. *Ectopia lentis*, typically inferonasal (Fig. 8.84), usually occurs by the age of 10 years. The zonule, which normally contains high levels of cysteine, disintegrates so that accommodation is often lost.
   b. *Secondary angle-closure glaucoma* may occur as a result of pupil block caused by lens incarceration in the pupil, or a total dislocation into the anterior chamber.
4. **Hyperlysinaemia** is a very rare, AR, inborn error of metabolism caused by a deficiency in lysine alpha-

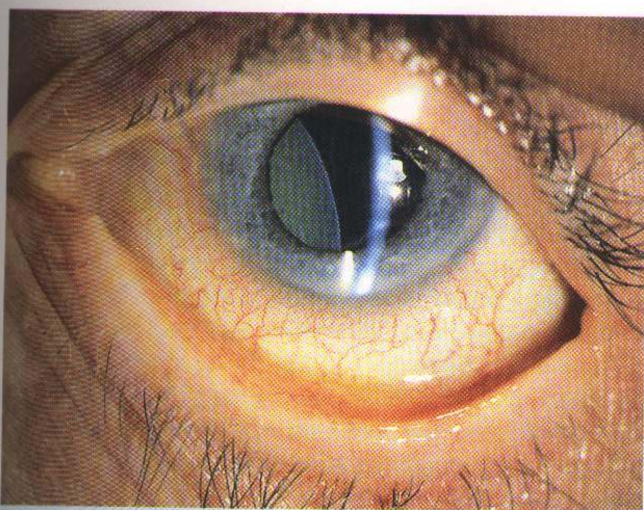


Fig. 8.84
Inferonasal lens subluxation in homocystinuria

ketoglutarate reductase. Systemic features include lax ligaments, hypotonic muscles, seizures and mental handicap. It is occasionally associated with ectopia lentis.

5. **Sulphite oxidase deficiency** is a very rare, AR disorder of sulphur metabolism characterized by progressive muscular rigidity, decerebrate posture, mental handicap and demise usually before the age of 5 years. Ectopia lentis is universal.
6. **Stickler syndrome** is occasionally associated with ectopia lentis, retinal detachment being the most common problem (*see* Chapter 12).
7. **Ehlers–Danlos syndrome** (*see* Chapter 20) is occasionally associated with ectopia lentis.

Management

The main complications of ectopia lentis are (a) refractive error (lenticular myopia), (b) optical distortion due to astigmatism and/or lens edge effect, (c) glaucoma and, rarely, (d) lens-induced uveitis.

1. **Spectacle correction** may correct astigmatism induced by lens tilt or edge effect in eyes with mild subluxation. Aphakic correction may also afford good visual results if a significant portion of the visual axis is aphakic in the undilated state.
2. **Surgical removal** of the lens, using closed intraocular microsurgical techniques, is indicated for cataract, lens-induced glaucoma, uveitis or endothelial touch.